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BEYOND THE DIAGNOSIS OF CYSTIC FIBROSIS : A STUDY OF DISEASE-RELATED KNOWLEDGE IN PATIENTS AND THEIR FAMILIES

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THESIS SUBMITTED FOR THE DEGREE

OF DOCTOR OF PHILOSOPHY

UNIVERSITY OF CAPE TOWN

JUNE 1988

ABSTRACT

A cross-sectional survey was conducted among 60 families with a child with cystic fibrosis (CF) to assess their medical knowledge of the illness, their subjective understanding of the patient's present and future health, how much they wanted to be told about the illness, their information needs and sources and to identify important sociodemographic and medical correlates of these assessments. The study also examined the relationship between knowledge of CF and personal and family adjustment.

The sample comprised all CF patients, within the Western Cape Hospital Region, who had attended the Red Cross War Memorial Children's Hospital in Cape Town over a two year period. Data were collected by means of structured questionnaires administered to 54 fathers, 60 mothers, 18 patients (aged 9-22 years) and 29 siblings (10-23 years). Respondents completed the questionnaires in their homes.

A 63 item multiple choice test, with acceptable psychometric properties, was used to assess medical knowledge. Parents and patients correctly answered approximately three-quarters, and siblings two-thirds, of the items. Mothers were slightly, yet significantly, better informed than fathers and siblings were significantly less informed than parents and patients. Family members were most knowledgeable about general CF facts, physiotherapy, gastrointestinal symptomatology and treatment and anatomy. They were less well-informed about respiratory symptomatology and treatment and nutrition. Parental knowledge of genetics and reproductive risks was mediocre, while that of patients and siblings was poor. Family members were least knowledgeable about general medical facts and terminology. Misconceptions, gaps and errors in CF knowledge, which could lead to inadvertent non-compliance, were identified.

Single item ratings of the patient's present and future health revealed highly positive perceptions among all respondents.

Index ratings of information preferences showed that most family members wanted complete and comprehensive information about CF. Measures of family members' information needs indicated that, in general, they wanted less information about symptomatology, treatment and genetics and more information about the psychosocial and future implications of CF. Fathers and siblings desired more medical information than mothers and patients.

Parents and patients relied most often on CF clinic doctors, their spouse/parents, prayer and faith and CF literature for information and guidance about the illness. Used less frequently were the CF Association and other parents with a CF child, while the hospital physiotherapists, social worker, dietician, the family doctor, local pharmacist and priest/minister were the least used sources. Siblings depended on mothers for their CF information. Based on index ratings of communication problems, most parents and patients did not experience difficulties when seeking information from the clinic doctors. Among a minority, the tendency to forget what they were told, a lack of privacy at the clinic and use of jargon presented problems. Parental index ratings of communication problems were inversely related to their CF knowledge.

Among parents, only social class was significantly associated with CF knowledge, perception of the patient's future health, magnitude of information needs, receipt of CF information, use of selected sources of information and the experience of communication problems with doctors. Information preferences were independent of social class. A similar social class effect was not noted for patients and siblings.

Parents who obtained higher scores on the test of CF knowledge and had lower information needs also reported less self-rated anxiety and depression.

Findings suggest the need for an ongoing assessment of parents' and patients' CF knowledge to identify areas of ignorance and to dispel misconceptions. The medical team needs in addition, to recognize that most family members want maximum information about CF and far more information about its psychosocial and future implications. The lower levels of CF knowledge and higher information needs of parents from the lower social classes warrant further consideration by health care professionals.

ACKNOWLEDGEMENTS

My sincere thanks are extended to Dr Chris Molteno, my supervisor, and to Dr Blanche de Wet for their ongoing encouragement and positive criticism. Dr De Wet is also thanked for translating the questionnaires into Afrikaans.

Thanks are due to Professor Malcolm Bowie and Drs Ivor Hill and Joe Ireland, of the Cystic Fibrosis Clinic, for their comments concerning the medical content of the questionnaires. Professor Eric Meyer of the Teaching Methods Unit of the University of Cape Town is thanked for his technical review of sections of the study questionnaires.

Special thanks are extended to the CF patients and their families who made the study possible.

Statistical analysis was provided by Dr Carl Lombard and Mrs Ria Laubsher of the Institute of Biostatistics of the Medical Research Council. I gratefully acknowledge their contribution.

I also extend my sincere thanks to Mrs Lynore Heuer for typing the thesis. I thank Mr Tony Rodrigues and Ms Jenny Altschuler for their help with the preparation of figures and drawings.

The study was supported by a grant from the Human Sciences Research Council.

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ABBREVIATIONS

Analysis of Variance	ANOVA
Chi-square	Chi^2
Coefficient of Determination	r^2
Cystic Fibrosis	CF
Degrees of Freedom	df
Depression Self Rating Scale	DSRS
Gastrointestinal	GIT
Hospital Depression and Anxiety Scale	HAD Scale
Life Orientation Test	LOT
Less than	$<$
Number (data base)	n
Probability	p
Revised Children's Manifest Anxiety Scale	RCMAS
Spearman Rank Correlation Coefficient	r_s
Standard Deviation	SD

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CHAPTER 1

INTRODUCTION

1.1 INTRODUCTION AND AIMS

Cystic Fibrosis (CF) is no longer a uniformly fatal childhood illness. Rapid advances in medical science have meant that increasing numbers of CF patients are reaching young adulthood (Mearns 1986). In order to cope with the acute and long-term demands of this progressive illness, parents and patients are faced with the ongoing task of accumulating and assimilating new and often complex medical information, some of which may be crucial to the quality of the child's survival. In addition to the medical facts, family members must learn about the psychosocial and future implications of this life-threatening illness.

For its part, the medical team has to decide what information about the illness CF families want and need to know. Yet a search of the CF literature indicated that scant attention had been given to the scientific measurement of either the disease-related knowledge or the information needs of CF families.

Although a section of the available evidence pointed to satisfactory levels of knowledge among CF patients and parents (Kulczycki, Robinson and Berg 1969; Tropauer, Franz and Dilgard 1970; Boyle, di Sant'Agnese, Sacks et al 1976; Bywater 1981 and Strauss and Wellisch 1982), due to the impressionistic and global nature of the assessments, it was not possible to accurately determine what they did or did not know. On the other hand, where research had focused on one specific aspect of knowledge, for example genetics, the findings suggested that adequate levels of comprehension were present in only a minority of CF parents (Leonard, Chase and Childs 1972; McCrae, Cull, Burton and Dodge

1973 and Burton 1975). A more recent study also noted that performance varied across the different domains of CF knowledge (Nolan, Desmond, Herlich and Hardy 1986). Using a standardized questionnaire, Nolan and his team (1986) found that parents and patients were well-informed about disease pathophysiology and treatment, that their grasp of genetics was fair but that their understanding of reproductive risks and male sterility was poor. Unfortunately small sample size limited the generalizability of these findings. No studies were located in the published literature which had examined the CF knowledge of siblings. Thus from the limited empirical findings it was not possible for the medical team to predict, with any degree of precision, likely levels, gaps or misconceptions in the CF knowledge of family members.

The need for a scientific assessment of family members' understanding of CF was further underscored by findings from equivalent research conducted among other groups of chronically ill children. These studies found that patients (Johnson, Pollak, Silverstein et al 1982; Beales, Holt, Keen and Mellor 1983 and Lorenz, Christensen and Pichert 1985) and parents (Kennel, Soroker, Thomas and Wasman 1968; Abdurrahman 1978; Reddihough, Landau, Jones and Rickards 1978; Jackson, Glasgow and Thom 1985 and Spykerboer, Donnelly and Thong 1986) were also ill-informed about many aspects of their illness. These studies too, it seemed, were plagued by such methodological shortcomings as small sample size, a lack of psychometrically sound measuring instruments and the failure to consider the multidimensional nature of disease-related knowledge. In addition, their findings concerning the effects of social and medical variables on parental and patient understanding of a disease, had produced inconclusive and often discrepant results.

Nor was it possible from existing research, to accurately determine what CF families wanted to know about the illness. Given the findings from research conducted among adolescent cancer patients that family members did not share the same information needs (Levenson, Copeland, Pfefferbaum and Silberberg 1983) and that discrepancies existed between the doctors and patients as to what each considered important information (Pfefferbaum, Levenson and Van Eys 1982), a detailed assessment of CF parents', patients' and siblings' medical and psychosocial information needs seemed essential.

A closely related issue, also with practical implications for information-giving, concerned how much CF patients and their families should be told about the illness. Whereas in the past doctors tended to withhold information, especially in the face of life-threatening illness, the current approach advocates an open policy towards the disclosure of information (reviews by Share 1972 and Van Dongen-Melman and Sanders-Woudstra 1986). Empirical CF research has not, however, established precisely how much families want to know about CF. And given the painful nature of some of the medical facts about the illness, it is conceivable that not all family members would have been equally desirous of being kept fully informed.

Moreover, in some CF samples the available evidence reflected a tendency to prefer less rather than more information. In two studies researchers encountered strong resistance among CF patients to knowing more about their illness (Boyle, di Sant'Agnese, Sacks et al 1976 and Bywater 1981), while Nolan and co-workers (1986) found that only one third of their patients wanted to know more about CF. In her study, Burton (1975) noted that slightly more than 50% of mothers and approximately 40% of fathers felt they would benefit from receiving more information about the disease. Falkman (1977), in turn, reported that even in the face of having received inadequate information at the time of diagnosis, almost 40% of her CF parents had not attempted to supplement their knowledge. Equivalent data were not available for siblings. This variation in responses across samples suggested the need for a broader and more reliable assessment of the information preferences of family members so that information-giving would accord with their expressed wishes.

Despite changing medical opinion towards the disclosure of sensitive information, uncertainty still surrounded the possible effect, on the psychological well-being of family members, of being presented with all the facts about a life-threatening illness. Indeed, it is only recently that paediatric researchers have begun to examine the relationship between knowledge of a disease and psychological adjustment.

Using standardized measures of anxiety and depression, Klein and Nimorwicz (1982) found a negative relationship between knowledge of haemophilia and self-reported psychological distress in family members. Susman, Hersh, Nannis et al (1982) reported that hospitalized cancer patients and their parents were able to retain an optimistic view of the future despite their knowledge that they might not have a future. Meanwhile in an earlier study among adult cancer patients, Cassileth and his team (1980) had also noted that despair was not an inevitable consequence of a thorough understanding of the illness. In the only available CF study to have treated disease-related knowledge as an explanatory variable, Pinkerton, Trauer, Duncan et al (1985) found that 'copers' were better informed about their illness than 'non-copers'. It was, however, not possible to determine how 'coping' or 'knowledge of CF' had been defined or measured. Thus while the available evidence suggested that knowledge of a chronic illness and emotional well-being could co-exist, the paucity and methodological shortcomings of this research made further investigation essential.

In the final analysis the collective data suggested that health care professionals were being called upon to educate and counsel CF families in the absence of reliable and valid data concerning their existing levels of CF knowledge, without clear guidelines as to the extent and nature of their information requirements and without knowing the possible psychological implications of a complete understanding of the illness. It seemed, therefore, that a study was needed which would accurately and comprehensively assess the disease-related knowledge, the information preferences and needs of family members, their main sources of information and which would also provide baseline data on the relationship between knowledge of CF and adjustment. These findings could then be used by the medical team to target their educational efforts by providing information relevant to the needs and present understanding of CF family members.

A sample of 60 CF families, who were attending the CF clinic at the Red Cross War Memorial Children's Hospital, was selected for the study. Data were collected by means of structured questionnaires administered to 54 fathers, 60 mothers, 18 patients and 29 siblings.

The study encompassed the following aims:

a. The assessment of current levels of medical knowledge about CF.
b. The assessment of perceptions (i.e. subjective knowledge) of the patient's present and future health.
c. The assessment of information preferences, that is, how much family members want to be told about CF.
d. The assessment of information needs concerning selected medical and psychosocial topics and whether this information had been provided in the past.
e. The identification and evaluation of sources of CF information and guidance used by family members.
f. The identification of important sociodemographic and medical correlates of the above measures.
g. The examination of the relationship between CF knowledge and the quantity and quality of informational support.
h. The examination of the relationship between CF knowledge and information needs and personal and family adjustment.

1.2 CHOICE OF THEORETICAL FRAMEWORK

A coping framework (Menaghan 1983) was selected for the present study. According to the coping model, people are rarely passive in the face of what happens to them (Folkman, Schaefer and Lazarus

1979). Coping has, in fact, been defined quite simply as the things people do to avoid being harmed by stress (Pearlin and Schooler 1978). These coping efforts may be intrapsychic (e.g. avoidance, denial) or behavioural (e.g. information-seeking) and represent the individual's attempts to manage the demands that tax or exceed his available resources (Folkman, Schaefer and Lazarus 1979).

Coping resources, on the other hand , refer not to what people do but to what is available to them in order to enhance their coping efforts (Pearlin and Schooler 1978). Psychological resources include an individual's personality and cognitive characteristics (e.g. optimism, knowledge) that help him to withstand stress (Menaghan 1983); social resources encompass the informational, material and emotional support provided by interpersonal networks such as the family and medical team (Moos and Billings 1982); while utilitarian resources (e.g. socioeconomic status) are likely to increase an individual's coping options (Folkman, Schaefer and Lazarus 1979).

The choice of a coping framework was motivated by several reasons. Firstly, a coping model discarded the pathology-based approach to CF research and focused, instead, on identifying factors which could facilitate management of the illness (Gayton, Friedman, Tavormina et al 1977; Drotar 1981 and Venters 1981). Intuitively, 'knowledge' of a disease seemed to be a prerequisite for successful coping, particularly where this included a knowledge of the treatment regimen.

Secondly, a coping model provided an appropriate framework for bridging the gap between research which in the past had treated 'knowledge' of a disease and the 'information needs' of families as separate research issues. Thus according to the coping model, knowledge could be conceptualized both as a coping resource which facilitates management and as an outcome of such well-described coping strategies as information-seeking (McCubbin, McCubbin, Patterson et al 1983).

Finally a coping model permitted the development and testing of hypotheses concerning the value of the various coping responses and resources. It was thus possible, for example, to evaluate the effect of social class membership on the amount of information given to family members.

1.3 OVERVIEW OF THE STUDY

The methodology of the study is presented in Chapter 2. Because the study encompassed a wide spectrum of aims, these are divided between two chapters: Chapters 3 and 4 each contain a literature study and the measures and findings pertinent to the particular aims of that chapter. Given the interdependence of results and to avoid repetition, the findings from both these chapters are discussed in Chapter 5. Practical recommendations based on these findings are also incorporated in the discussion. A summary of the major findings and recommendations for future research are presented in the final chapter.

CHAPTER 2

METHODOLOGY

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2.1 CHOICE OF RESEARCH DESIGN

The present study used the cross-sectional survey method to meet its aims which were both exploratory and analytical in nature. The primary function of this type of survey is to provide accurate, quantitative - descriptive data which can be generalized to a designated population (Tripodi 1981). This method is particularly suited to collecting factual data as well as psychological information such as opinions and attitudes (Kerlinger 1975).

Despite criticism that the survey method is inflexible, superficial and lacking in longitudinal dimension (Weiss 1966), it was particularly suitable for the present study for several reasons. Since four family sub-groupings (fathers, mothers, older patients and siblings) were to be assessed separately the more standardized approach of the survey would facilitate both within-and-between group comparisons. The inclusion of two language groups (English and Afrikaans) also necessitated a structured approach which would enable accurate and uniform translation of all measures prior to the study. It was felt, furthermore, that the inclusion of families with children of varying developmental levels would provide a measure of longitudinal perspective when interpreting the data (Darling 1979).

2.2 THE STUDY SAMPLE

The study sample comprised a complete, consecutive series of all patients, with a confirmed diagnosis of CF, and their families who had attended the CF clinic at the Red Cross War

Memorial Children's Hospital in Cape Town, between January 1, 1984 and December 31, 1985. To facilitate data collection, families who were not resident in the Western Cape Hospital Region (as designated in the Report of the Director: Hospital Services 1984) were excluded from the sample.

According to hospital computer records, 63 families met these criteria. During the course of the study one child was rediagnosed as NOT having CF which reduced the total number of eligible families to sixty two.

2.2.1 Representativeness of the Sample

The sample included ALL CF patients, within the Western Cape Hospital Region, who had attended the Red Cross War Memorial Children's Hospital during the prescribed sampling period. Since this Hospital is the only referral centre for CF patients in this region (Hill, Macdonald, Bowie and Ireland 1988) it was assumed that the CF clinic included most known patients with CF.

2.3 TECHNIQUES OF DATA COLLECTION

2.3.1 Procedures

Data collection took place over a period of 3 months (July-September 1986). Due to an incomplete address in the hospital file, one rural family could not be traced. This patient was a known clinic defaulter and had attended clinic only twice during the two year sampling period. All parents, in the remaining 61 families, were informed of the explicit statement of the theoretical and practical importance of the study as well as a guarantee of complete confidentiality. Patients and siblings who were 12 years and older were also invited, with parental permission, to take part in the study.

Shortly thereafter, a follow-up telephone call was made to each family to find out whether or not they were prepared to take part in the study. Although no family had refused to take part, one out-of-town family failed to return their completed questionnaires in time for analysis. All parents allowed, indeed welcomed, their older children's participation. In 7 families, parents expressly requested that patients (n = 5) and siblings (n = 3) who were less than 12 years also be allowed to answer the questionnaires. Their ages ranged from 9 to 11 years.

Questionnaires were personally delivered to families in their homes. Where families lived more than 100 kilometers from Cape Town, questionnaires were delivered during their visits to the CF clinic. Within 3 to 4 weeks of delivery, families were again contacted telephonically to establish whether the questionnaires had been completed. These were personally collected during follow-up home visits. Four sets of questionnaires were returned by post.

This personalized approach to data collection resulted in 60 out of the 61 sets of questionnaires being returned - a response rate of 98.36 per cent (or 96.78% of the original sample of 62 families). Questionnaires were completed by 60 mothers (6 of whom were single parents), 54 fathers, 18 patients and 29 siblings. A complete sociodemographic and medical profile of the final sample is presented in Tables 3.4 and 3.5 in Chapter 3.

In 4 families the author had to re-contact members regarding missing data on their questionnaires.

As a token of thanks, each family was furnished with a copy of relevant preliminary results from the study. Included in each report was a copy of the correct answers to the medical questions. It was hoped that by providing feedback, the assessment of families' knowledge about CF would become a teaching tool in itself (Dunn, Bryson, Hoskins et al 1984).

2.3.2 Questionnaires

Data were collected by means of structured self-administered questionnaires. Three separate questionnaires were designed for parents, patients and siblings (Appendices 1 to 3). The format and content of these questionnaires were very similar, with differences relating typically to the perspective of the respondent (for example, it is my right to know everything about my condition/about my child's condition/about my brother or sister's condition) and to the appropriateness of selected measures for a particular family grouping. The format of the questionnaires for fathers and mothers was the same.

In order to meet the wide spectrum of aims encompassed by the present study, a variety of measures was used. These included measures of family members' knowledge about CF, the degree to which they preferred to be kept informed about the illness, how much they wanted to know about selected medical and psychosocial topics, whether or not they had been given this information before and their utilization and evaluation of potential sources of informational support. Also included were standardized measures of anxiety, depression, family functioning and dispositional optimism. The purpose, design and psychometric properties of individual items and scales are described in Chapters 3 and 4. The location of these measures in the original questionnaires, according to family grouping, is presented in Appendices 4 and 5.

Because the questionnaires included a number of measures, particular attention was paid to lay-out and to easing the burden of response. Most answers, therefore, required only a circle, a tick or a short answer. The few open-ended questions were included to give respondents the opportunity to freely express and expand upon their opinions.

Despite their importance, sensitive topics which had the potential for creating undue anxiety, such as those explicitly relating to sterility, possible complications or shortened lifespan, were excluded from the study. This step was necessary because the author could not be sure whether these issues had ever been discussed with the families.

All questionnaires were translated into Afrikaans by a senior colleague. They were subjected to reverse translation by a second colleague to ensure that the meaning of the items had remained unchanged during the translation process.

It was not possible to conduct a pilot study of the questionnaires as ALL families within the Western Cape Hospital Region who were attending the CF clinic, were included in the main study. The questionnaires were, however, repeatedly given to colleagues for review and criticism during the developmental stages. They were also approved by the Institute of Biostatistics of the South African Medical Research Council.

2.3.2.1 Reading Levels of the English Questionnaires

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The reading levels of the English questionnaires were assessed according to the Flesch Formula (Spadaro, Robinson and Smith 1980). Readability formulas are designed to make quick and easy assessments of the complexity of written material. The most widely used is the Flesch score which combines words per sentence (as a measure of sentence complexity) and syllables per 100 words (as a measure of vocabulary difficulty) to provide a numerical Flesch reading ease score. A high score (over 69) indicates an easy reading level, while lower scores indicate more difficult levels (Nicoll and Harrison 1984).

An average Flesch score was calculated from two samples of written material from pages 1 and 12 of the three sets of English questionnaires (Appendix 6). The Afrikaans questionnaires were not included in this analysis as the Flesch formula has not been validated on the Afrikaans language.

A summary of the Flesch scores and their meaning is presented in Table 2.1.

TABLE 2.1

INTERPRETATION OF FLESCH READING EASE SCORES FOR ENGLISH QUESTIONNAIRES ACCORDING TO FAMILY GROUPING

FAMILY GROUPING	FLESCH SCORE	DESCRIPTION OF READING LEVEL	GRADE READING LEVEL	
Parents	68.17	Standard	8 - 9	(Std 6-7)
Patients	78.94	Fairly easy	7	(Std 5)
Siblings	88.72	Easy	6	(Std 4)

These scores indicate that the English questionnaires were written within acceptable levels of readability. (The mean number of completed years of education for patients and siblings in the present sample was 7.67 and 8.6 years respectively).

It is necessary to point out that initially the author had intended to administer the questionnaires to children 12 years and older. Children from as young as 9 years were included on parental request. Thus, for these younger respondents the reading levels may well have been too advanced (mean number of completed years of education for patients and siblings under 12 years, was 4.2 and 3.67 respectively).

2.4 STATISTICAL ANALYSIS

Statistical analysis was undertaken by the Institute of Biostatistics of the South African Medical Research Council. Raw data were coded by the author and then entered into the computer at the Institute. The BMDP (Biomedical Data Processing) and SAS (Statistical Analysis System) Statistical Packages were used for these analyses (Dixon, Brown, Engelman et al 1981 and SAS Institute Inc. 1985). Since the variables used in the study were either categorical or ordinal, the most commonly used statistical procedures were non-parametric. These included Chi-square analysis, the Wilcoxon Sign Rank Test and Spearman Rank Order Correlations. The McNemar Test of Symmetry was the Chi-square test most frequently used for the matched pair analysis of categorical variables.

Analysis of variance and regression analysis were also selectively used in the study. These parametric techniques are sufficiently robust to withstand contravention of their basic underlying assumptions particularly regarding their requirement for interval scale measurement (Kohout and Norwood 1981). Furthermore, with few exceptions parametric tests were used only for the analysis of data from samples of fathers and mothers because of their larger numbers. Univariate and multivariate analyses were used to establish the main or interaction effects of single and combined independent variables on a dependent variable. When a significant F value was noted in an analysis of variance, further pairwise testing using Gabriel's Test, was undertaken to determine which between group differences had contributed to the significant finding. Gabriel's Test is designed to control for an inflated Type 1 error rate. In order to establish the magnitude of a significant effect of an independent on a dependent variable, the r^2 statistic (the coefficient of determination) was calculated. This statistic is able to determine the proportion of variance in the dependent variable explained by the independent variable (Kohout and Norwood 1981). The multiple coefficient of determination, R^2, was calculated for the multivariate model and is analogous to the bivariate r^2 statistic.

Probability levels of 0.05 were used for most analyses, with the exception of Spearman correlations where a more stringent level (0.005) was selected to minimize chance findings. Similar significance levels (0.005) were used when analyses entailed a large number of ANOVAS. Probability levels have been reported in the text only when they reflected statistically significant results. Where non-significant p values were reported, they have been tabulated in the Appendices. Unless otherwise stated, degrees of freedom equal 1 for the Chi-square tests and for the analysis of variance.

The reliability, or internal consistency, of the measures used in the study, was determined by Cronbach's alpha (Kerlinger

1975). Since Cronbach's alpha is, in part, a function of the number of items in a measure (Nelson 1981), wherever possible single item scores were summated to form index (i.e. composite) scores (Babbie 1975). These were then used as dependent or independent variables depending on the context of the analysis.

CHAPTER 3

KNOWLEDGE ABOUT CYSTIC FIBROSIS AMONG FAMILY MEMBERS

3.1 INTRODUCTION

This chapter is concerned with meeting the following aims of the study:

a. The assessment of current levels of medical knowledge about CF.

b. The assessment of perceptions of the patient's present and future health.

c. The identification of important sociodemographic and medical correlates of the above measures. The chapter begins with a review of the pertinent literature.

3.2 REVIEW OF THE LITERATURE

A noteworthy feature of most of this review has been the limited number of CF studies directly applicable to the measurement of family members' understanding of the illness. It has, therefore, been necessary to draw on the available evidence from equivalent research in other fields of chronic paediatric illness.

By way of an introduction to the chronically ill child's thinking about medical matters, the review begins by outlining selected findings from research which has investigated how children, both healthy and sick, develop a knowledge of their bodies, their health, physical illness and death. This is followed by a synopsis and evaluation of studies, across disease categories, which have assessed levels of knowledge in chronically ill children and their parents. The literature study is concluded with a review of paediatric research which has examined the link between disease-related knowledge and compliance in chronic illness.

3.2.1 Knowledge of Medical Matters in Sick and Healthy Children

A great deal of recent research has been devoted to investigating what well and sick children understand about medical matters (Maddux, Roberts, Sledden and Wright 1986), the professed goal of much of this research being to improve communication with sick children (Eiser 1985). In general, this research has been conducted within a framework of either chronological age or cognitive-developmental theory (Burbach and Peterson 1986). With the possible exception of the limited research into CF children's understanding of death, similar research has not been conducted amongst samples of CF children, and the author has, therefore, included equivalent studies from other chronic illness groups.

3.2.1.1 What Children Understand about their Bodies

Clearly what children - both healthy and chronically ill - understand about illness and treatment, must be influenced by their ideas about the content and functioning of the human body. A number of studies have explored what healthy children understand about their bodies (Tait and Ascher 1955, Porter 1974, Gellert 1978 and Eiser and Patterson 1983). The most commonly employed methodology was to ask children to draw the inside of their bodies and to answer questions relating to body parts and their functions. These studies found that the number of body parts mentioned by the children increased according to the age of the child. By the age of 7, most of the children mentioned the brain, bones, heart and blood. By the age of 10 to 12 years, they added the stomach and lungs and it was only during adolescence that they were likely to include the kidneys, liver or reproductive organs.

Knowledge of body function was also strongly related to the age of the child. At the least mature level, children were unable to differentiate between the body parts and their various functions (Eiser 1983). Six to 7 year olds recognized that a body part had a function, but their explanations tended to be very simple, for example, the heart is for loving or the brain is for thinking.

From 10 years upwards, children were able to recognize that the different organs had different functions. Thus before about 10 years, Gellert (1978) found that the children in her sample were unable to relate breathing to the lungs. Even in the older group, knowledge of the function of the liver and bladder was very sparse. Only 2 children (whose ages were not mentioned) in Eiser and Patterson's study (1983) could say that the liver was involved in digestion. These authors concluded that explanations of body function tended to focus on the external and visible aspects, while at the same time reflecting considerable confusion about what was taking place inside.

These same authors also found much ignorance regarding children's ideas about the various body processes. For example, most 6 year olds thought that food remained in the stomach forever, while at the 12 year old level only a very few children realized that food was converted and released into the blood stream. Knowledge of digestion was equally inadequate in Gellert's sample (1978). Between 8 and 9 years, children began to connect the digestive and excretory systems (Eiser and Patterson 1983). Below this age, the children either did not know what happened to food or else they believed it continued to move around the body in its original form (Gellert 1978).

① // In summary, the development of the child's knowledge of the body takes place over 3 age-related stages (Eiser 1985). The first stage, up to 7 years, is characterized by immature, global thinking about the body. During this period children confuse internal and external aspects of the body and they are unable to differentiate between the parts of the body and their functions. Importantly, the very young child is unable to reason beyond appearance, that is, beyond what he can see on the outside happening to his body. Between 7 and 11 years, children's knowledge of the number of body parts increases but they are unable to relate these various parts of the body together. By adolescence, responses are more mature and children are able to identify body parts, their function as well as the workings of the various systems such as circulation and digestion. These 3 stages roughly correspond to the Piagetian stages of preoperational, concrete operational and formal operational thought.

It is worth noting that ignorance of the body and its functions is not isolated to children. Several studies have highlighted equally inadequate and inaccurate knowledge of the body among adult samples (Tait and Ascher 1955; Pool 1980 and Pearson and Dudley 1982). Accompanying this lack of knowledge were some startling misconceptions, examples of which include one woman who confused her stomach and uterus and one man who believed he had two stomachs (Pearson and Dudley 1982). These authors concluded that this lack of knowledge was a major stumbling block in the way of promoting patient understanding of their condition.

A few studies have also examined chronically ill children's understanding of their bodies. Eiser, Patterson and Tripp (1984a) found little difference between a sample of diabetic children and a healthy control group in respect of their knowledge about their bodies. Indeed, they found no differences between the groups in either the number of internal body parts mentioned or in their explanation of the function of selected body parts. One noteworthy, but anticipated, finding was that the diabetic children were more likely to include a pancreas in their drawings. The authors interpreted this latter finding as possibly reflecting a difference in attitude, rather than knowledge, toward their bodies resulting in the diabetic children attributing more importance to some of their body parts compared to healthy children. A similar finding was reported by Korsch, Negrete, Gardner et al (1973) in a sample of children with chronic renal disease. These authors noted a tendency among these children to include detailed patterns of arteries and veins in their drawings, possibly reflecting their exaggerated experience with shunt and intravenous infusion procedures.

Beales, Holt, Keen and Mellor (1983) explored the understanding of 75 children, aged 7-17, who were suffering from juvenile chronic arthritis. In keeping with the Piagetian framework, they found distinct differences in the beliefs and concepts of children aged 7 to 11 and those aged 12 to 17 years. The younger group exhibited very little understanding of internal pathology and described their disease in terms of outward, largely visible manifestations such as stiff joints and sore fingers. Their images of what was taking place below the surface of their skin were very vague. This contrasted sharply with the responses of the older group who were far more inclined to relate their symptoms to underlying pathology. Thus bones and blood figured prominently in the imagery. Much of their imagery, however, was oversimplified and reflected a tendency to exaggerate the extent of internal damage. A similar finding was reported by Kaufman and Hersher (1971) who noted that their sample of diabetic children consistently visualized the insides of their bodies as being bad and abnormal.

Beales and his co-workers (1983) also explored the implications of bodily perceptions on their children's understanding of their different therapies. They found that children who had a greater understanding of the internal manifestations of their disease, also had a better appreciation of the purposes of their therapy. Whereas the younger group of children was unable to see how tablets or injections could possibly have an effect on their joints, the older group at least appreciated that there must be a physical connection within the body whereby ingested medicines could reach the affected joints. This is one of the few studies which has attempted to link knowledge of body parts and functions to an outcome variable such as compliance.

3.2.1.2 What Children Understand about Physical Illness, Health, Treatment and the Prevention of Illness

Evidence is accumulating that a child's cognitive development is particularly important in his[1] knowledge of physical illness (Carandang, Folkins, Hines and Steward 1979; Bibace and Walsh 1980; Perrin and Gerrity 1981; Brewster 1982), contagion (Kister and Patterson 1980), healing and recovery (Neuhauser, Amsterdam, Hines and Steward 1978) and prevention (Blum and Stark 1985). Yet other studies indicate how children's increasing age leads to qualitatively improved understanding of illness (Brodie 1974; Campbell 1975; Millstein, Adler and Irwin 1981; Eiser, Patterson and Eiser 1983; Eiser, Patterson and Tripp 1984b), health (Natapoff 1978) and prevention (Eiser, Patterson and Eiser 1983).

Understanding Physical Illness

Recent review articles by Pidgeon (1985) and Burbach and Peterson (1986) have respectively evaluated the roles of chronological age and cognitive development in the progression of healthy and sick children's understanding of physical illness.

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[1] For ease of reading, only the male pronoun has been used throughout the text.

With increasing age and cognitive development these authors found that:

i. Children's conceptualization of illness moves from being global and non-specific to being specific and disease-orientated.

ii. Children come to associate illness with infection and germs rather than punishment.

iii. Children come to appreciate the role of psychosocial and emotional factors in the aetiology of illness.

iv. Children come to understand the concept of contagion.

v. Children begin to feel that they have more control over illness and recovery.

As yet research has not reached any definite conclusions regarding the contribution which illness experience and hospitalization make to a child's understanding of physical illness (Eiser 1985). According to the Piagetian framework, a chronically ill child should have a more mature understanding by virtue of his experience with medical matters, but the current research findings are inconsistent. Some researchers (Brewster 1982; Eiser, Patterson and Tripp 1984b; Carracio, McCormick and Weller 1987) have found no differences between the illness concepts of healthy children and their chronically ill or hospitalized counterparts, while others report less mature concepts (Carandang, Folkins, Hines and Steward 1979). In Campbell's study (1975) it was only the older children with poorer health, who had more sophisticated illness concepts than their healthier peers. The younger, unhealthy children (less than 9.6 years) on the other hand evidenced the most unsophisticated illness concepts of both healthy and sick children. This finding, together with the tentative findings of Redpath and Rogers (1984), suggest that the stress associated with illness and hospitalization may have a more detrimental effect on the younger rather than older child's understanding of illness. Further research, controlling for variables such as age and illness characteristics, is called for. Similarly research needs to explore the impact of stress (such as might be caused by chronic illness) on a child's conceptual abilities and ideas about illness.

Understanding Health and the Prevention of Illness

The development of sick and healthy children's understanding of health follows a pattern similar to their understanding of illness. Children move from a sense that their health is vulnerable to external influences to a realization that their health is dependent on their own behaviour (Bibace and Walsh 1980). Most studies (Natapoff 1978; Millstein, Adler and Irwin 1981 and Eiser, Patterson and Eiser 1983) show that children interpret health positively to mean that they are able to take part in desired activities such as playing with friends, playing sport and generally being energetic. Furthermore, when Eiser, Patterson and Tripp (1984b) compared diabetic and healthy children's concepts of health, they found no significant differences. They interpreted this to mean that chronic illness does not seem to affect a child's definition of health.

In order to stay healthy, children stressed the importance of diet (Eiser, Patterson and Eiser 1983). With increasing age, children were able to say why certain foods were likely to improve health, while others were detrimental. 'Bad' foods were identified as those containing too much sugar and grease. In general, however, these authors did not find that their sample (6-11 years) had an adequate understanding of prevention.

Blum and Stark (1985) make the point that 'prevention involves taking or avoiding an action to improve one's health or diminish one's risk of illness in the future' (p.27) and, as such, it necessitates an understanding of future time which, in turn, really only develops in adolescence. They also stress how crucial it is for the chronically ill adolescent to understand prevention so as to forestall exacerbations of their illness. They do caution, however, that before an adolescent is likely to invest in the future through preventive behaviour, he must feel 'the future is worth investing in' (p.28). This has obvious implications for the adolescent with a life-threatening illness such as cystic fibrosis.

Understanding Medical Treatment

The role of cognitive development has also been implicated in the child's understanding of treatment (Brewster 1982 and Beales, Holt, Keen and Mellor 1983) of hospitals, operations and the role of medical personnel (Redpath and Roger 1984) and of medical equipment (Steward and Regalbuto 1975). Age has been shown to influence the child's understanding of hospitalization (Eiser and Patterson 1984) and of the roles of doctors and patients (Haight, Black and DiMatteo 1985). Eiser (1985b) has recently reviewed the literature pertaining to the child's understanding of treatment and medical procedures. She makes the point that almost without exception, young children interpret treatment to be a form of punishment. Furthermore, they frequently misperceive the reasons for different medical procedures. It is only from about 10 years onwards, that children are likely to recognize the potential long-term benefits of treatment (Brewster 1982).

Although the studies reviewed so far provide valuable insight into children's illness concepts, their findings must be treated with caution. In their review article, Burbach and Peterson (1986) summarized the major methdological shortcomings of much of this research as follows: poorly described samples and measuring instruments, observer bias, failure to control for potentially confounding variables such as social class or illness severity and the absence of reliability and validity data.

In summary, what sick and well children understand about physical illness, health, treatment and prevention is largely a function of their cognitive development and chronological age. This understanding progresses from the global, somewhat magical, thinking of the preoperational child (2 to 7 years) through the concrete and more logical thinking of the concrete operational child (7 to 11 years) to the sophisticated, abstract conceptualization of the adolescent.

The following section will review relevant research which looks at how the healthy and sick child develops an understanding of death.

3.2.1.3 What Healthy and Chronically Ill Children Understand About Death

Similarly to his conceptions of physical illness and its treatment, the child's understanding of death becomes increasingly more sophisticated and realistic with increasing age (Kane 1979) and cognitive development (Nagy 1948; Koocher 1973; White, Elsom and Prawat 1978 and Kane 1979). While the egocentric 3 year old is likely to view death as an event that happens to other people but not to himself, by 8 or 9 years of age most children have developed mature death concepts (Kane 1979 and Reilly, Hasazi and Bond 1983); that is, death is viewed as an internal process involving the cessation of bodily functions as well as being universal, permanent and final (Kane 1979). A recent study (Lansdown and Benjamin 1985) found that healthy children from as young as 5 years old have a good grasp of the meaning of 'dead'.

In a similar vein, research has shown that fatally ill children from as young as 6 years old can comprehend the meaning of death and dying (Waechter 1971 and Spinetta, Rigler and Karon 1973). With the exception of Waechter (1971) who included 6 CF children in her sample, most of the literature pertaining to the CF child's understanding or awareness of death is anecdotal, based on unconfirmed clinical experience (Patterson, Denning and Kutscher 1973) and interview data. For example, Gratzick (1973) using his personal and group experience with 10 hospitalized CF patients, aged 6-26, outlined their understanding of the fatal nature of their illness within an age-related framework. Although he felt that a primary concern of the hospitalized CF child was the possibility of dying, he did not believe that the 6 to 7 year old CF child equated his disease with death. The 8 to 10 year old CF child, however, seemed to equate death and illness in a more personal way, usually as a result of experiencing the death of another CF child. This age group also tended to rationalize that those CF children who died had been sicker than they were. Although the 11 to 14 year olds seemed to have a good understanding of death, Gratzick (1973) found that they were more oriented towards life and coped with the possibility of their own death through repression. From 15 years upwards, he reports that the CF children had a comprehensive understanding of their personal mortality although the topic was seldom openly discussed. Unfortunately, these interpretations were not supported with any empirical evidence.

Other researchers (Lawler, Nakielny and Wright 1966; McCollum and Gibson 1970; Tropauer, Franz and Dilgard 1970; Burton 1975; Boyle, di Sant'Agnese, Sack, Millican and Kulczycki 1976) have also found a heightened preoccupation with death in their CF samples, although the express purpose of their studies was not the assessment of these children's understanding of death. The age at which this awareness developed, varied between samples. Burton (1975), for example, noted that a precocious awareness of death developed between 2 and 6 years of age, often in relation to treatment. These youngsters would openly ask their mothers if they would die were they to discontinue their therapy. On the other hand, mothers in the McCollum and Gibson (1970) study reported a growing consciousness of the terminal nature of CF in their children aged from between 8 and 12 years.

Yet other studies (Kane 1979 and Reilly, Hasazi and Bond 1983) have explored the role of experiential events, such as the death of a significant other, as a basis for promoting an understanding of death. These authors found that the effect of death-related experiences in promoting a precocious understanding of death, was most marked in the 5 to 6 year old age group.

The important role that experience plays in promoting the fatally ill child's understanding of death is well described by Bluebond-Langner (1977). Although her sample included only leukaemic children her findings are relevant to other chronically ill children. She proposed that the fatally ill child's understanding of death develops as he passes through various stages such as diagnosis, remission and relapse. She found that the death of another child with a similar illness, was particularly significant in determining the fatally ill child's realization that he too might die from his illness, irrespective of his age or cognitive development. To quote from her findings: 'there are 3 and 4 year olds of average intelligence who know more about their prognosis than very intelligent 9 year olds. The reason for this is that the 9 year olds may still be in their first remission, have had fewer clinic visits, and hence less experience' (1977, p.54).

Clearly, therefore, illness-related experiences must also have been partly instrumental in promoting an early understanding of death in the samples of Waechter (1971) and Spinetta, Rigler and Karon (1973) as most of the parents claimed that they had not told their children of the fatal nature of their illness. More specifically, several CF studies have also mentioned how inopportune comments during ward rounds (De Wet 1984), insensitive comments of siblings or peers (McCollum and Gibson 1970 and Burton 1975), sensational newspaper or magazine articles or well-publicised fund raising campaigns in the mass media (McCollum and Gibson 1970) contribute to the fatally ill child's heightened awareness of the terminal nature of his illness.

Orbach, Gross, Glaubman and Berman (1985) do not, however, feel that a child's acquisition of knowledge about death is simply linear and accumulative. They maintain that emotional factors may also influence the child's ideas about death. They present findings, from a sample of healthy children, which suggest that anxiety may interfere with and distort a child's understanding of death at certain stages of his development. Furthermore, these authors stress the importance, for therapeutic and educational purposes, of qualitatively distinguishing between immature death concepts which result from cognitive immaturity and those which are caused by defensiveness. This distinction has particular relevance for fatally ill children as research has already identified high levels of death-related anxiety in these children (Waechter 1971; Spinetta, Rigler and Karon 1973 and Spinetta and Maloney 1975).

In summary, research shows that healthy and chronically ill children can comprehend the meaning of death and dying from about 5 years of age. Indeed, some research has found that in fatally ill children, this awareness may develop from an even earlier age due largely to their exposure to illness-related experiences. Although not based on rigorous research, available evidence does seem to suggest that CF children gain an understanding of the incurable nature of their illness, together with the possibility of a limited lifespan, from an equally early age. Empirical research is, nonetheless, needed to evaluate the role of cognitive, experiential and emotional factors in the development of the CF child's understanding of death.

3.2.2 Knowledge of Chronic Illness in Patients and Parents

It is necessary to point out that this section is not intended to provide a content analysis of what chronically ill children do or do not know about their illnesses. Instead its purpose is to synthesize relevant literature so as to provide a broad perspective of the main correlates of this knowledge and to highlight the methodological shortcomings and problems associated with this field of research. Analogous findings from studies which have measured parental knowledge are also reviewed in this section. As already noted equivalent CF research is limited.

A chronological summary of empirical studies which have examined chronically ill children and their parent's knowledge of their illnesses is presented in Table 3.1 with respect to

TABLE 3.1

KNOWLEDGE OF CHRONIC ILLNESS IN CHILDREN AND THEIR PARENTS : A SYNOPSIS OF RESEARCH STUDIES

REFERENCE	SAMPLE CHARACTERISTICS			SAMPLING METHODS	MEASURING INSTRUMENT
	ILLNESS	SIZE	AGE OF CHILDREN		
Etzwiler (1962)	Juvenile diabetes	74 children	6-17	Attenders at a diabetic camp.	15-item multiple choice questionnaire. No psychometric data.
Maxwell and Gane (1962)	Congenital heart disease	150 families	Not given	Attenders at a cardiac clinic at a University Hospital.	Knowledge of disease explored during an interview. No details provided.
① Kennell, Soroker, Thomas and Wasmann (1969)	Rheumatic fever	60 parents	Under 15	Attenders at a rheumatic fever clinic.	Questionnaire - no details provided. Standard questions were asked but answers were open-ended. No psychometric data.
Taylor and Merrill (1970)	Duchenne muscular dystrophy	21 families	Not given	Attenders at a muscular dystrophy clinic.	Semi-structured interview - no details provided. No psychometric data.
(2) Collier and Etzwiler (1971)	Juvenile diabetes	129 children 141 parents	Not given	Random sample of diabetic children attending local junior and secondary schools in a prescribed area.	34-item multiple choice test - 'Diabetic Knowledge Test'. No psychometric data.

Continued on next page

Table 3.1 Continued

REFERENCE	SAMPLE CHARACTERISTICS			SAMPLING METHODS	MEASURING INSTRUMENT
	ILLNESS	SIZE	AGE OF CHILDREN		
Sibinga and Friedman (1971)	Phenylketonuria	79 parents (42 families)	Not given	Not stated, other than families attending a state-supported PKU programme.	An essay type questionnaire concerning the genetics, causation, symptoms, prognosis and treatment of PKU. Responses scored against an 'ideal' answer and for distortion. No data on reliability or validity.
Leonard, Chase and Childs (1972)	Cystic fibrosis, PKU and Down's syndrome	CF = 39 PKU = 7 Down's = 15	Not given	Non-random, drawn from specialist clinics.	Semi-structured interview and questionnaire on general and specific knowledge of genetics and probability. No reliability or validity data.
Partridge, Garner, Thompson and Cherry (1972)	Juvenile diabetes	54 children	14-18	Attenders from 'the same practice' - no further details.	25-item diabetic information test. Considered content validity.

Continued on next page

Table 3.1 continued

REFERENCE	SAMPLE CHARACTERISTICS			SAMPLING METHODS	MEASURING INSTRUMENT
	ILLNESS	SIZE	AGE OF CHILDREN		
Reiss and Menashe (1972)	Congenital heart disease	39 couples	Not given	Non-random attenders at routine follow-up heart clinics at a university hospital.	Ten minute interview during which parents were asked 5 standard questions. 35 couples were re-tested on 2 questions from one to four months later.
McCrae, Cull, Burton and Dodge (1973)	Cystic fibrosis	100 families	Not given	Not described.	Interview - no further details. Comprehension was rated as excellent, moderate or poor.
Burton (1975)	Cystic fibrosis	53 families (97 parents)	7 months - 16 years	All known CF families in Northern Ireland.	During a home-based interview, parents asked one question concerning the chances of their children's children having CF. Understanding graded as excellent, moderate or poor. No reliability or validity data.
McLucas, Hsia and Mick (1975)	Duchenne muscular dystrophy	26 mothers	Not given	Not given - high non-response rate.	Questionnaire and interview - no further details. No definition of adequate understanding.

Continued on next page

Table 3.1 continued

REFERENCE	SAMPLE CHARACTERISTICS			SAMPLING METHODS	MEASURING INSTRUMENT
	ILLNESS	SIZE	AGE OF CHILDREN		
Kupst, Dresser, Schulman and Paul (1976)	Congenital heart disease	88 parents	Under 14	Randomly selected attenders at a cardiac out-patient clinic.	Standardized interview covering 9 areas of knowledge. Recall assessed on a 4-point scale immediately following an interview and 4 weeks later.
Ludvigsson (1977)	Juvenile diabetes	58 children	6-17	Attenders at a specialist diabetic clinic at a university hospital.	30-item, written knowledge test. Administered under controlled conditions with no access to literature. 'Adequate knowledge' = 75% of maximum possible points. No psychometric data.
Abdurrahman (1978)	Various: Sickle cell disease TB Protein calorie Malnutrition Heart disease Malignancy More than one disease Miscellaneous	100 parents 40 13 13 5 5 3 13	Not given	Systematic sampling of every third patient admitted to a hospital ward or seen at OPD, fulfilling certain inclusionary criteria.	Questionnaire containing open and closed questions. Reliability assessed by retesting 12 randomly chosen parents.No criteria for 'adequate' understanding.

Continued on next page

Table 3.1 continued

REFERENCE	SAMPLE CHARACTERISTICS			SAMPLING METHODS	MEASURING INSTRUMENT
	ILLNESS	SIZE	AGE OF CHILDREN		
* Reddihough, Landau, Jones and Rickards (1978)	Asthma	41 families	9-11	Attenders at a specialist clinic & general medical clinics.	Questionnaire - open and closed questions. No psychometric data.
Ferencz, Wiegmann and Dunning (1980)	Congenital heart disease	73 children	14-25	All patients registered at 2 cardiac clinics. High non-response rate.	Questionnaire. No details of validity and reliability. Responses categorized as all correct, partly correct or none correct.
Hamburg and Inoff (1982)	Juvenile diabetes	211 children	5-19	Attenders at a diabetic camp.	20-item multiple choice questionnaire. Reliability assessed.
(5) Johnson, Pollak, Silverstein et al (1982)	Juvenile diabetes	151 children 179 parents	6-16	Not stated.	Multiple choice questionnaire - included general and problem solving questions. Reliability and content validity assessed. Included only items which were correctly answered by 2 physicians and a nurse.

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Table 3.1 continued

REFERENCE	SAMPLE CHARACTERISTICS			SAMPLING METHODS	MEASURING INSTRUMENT
	ILLNESS	SIZE	AGE OF CHILDREN		
Klein and Nimorwicz (1982)	Haemophilia	12 families (pilot study)	8-23	Attenders at a haemophilia clinic. Must have attended an educational programme.	58 true-false questions. No psychometric data.
Susman, Hersh, Nannis et al (1982)	Cancer	16 patient/ mother pairs	8-21	Purposive sample of hospital in-patients.	Structured interview. Knowledge assessed by trained interviewers on scales of 1 to 9. They were blind to the purpose of the interviews. Intraclass correlations between interviewers ranged between 0.98 & 0.67.
Beales, Holt, Keen and Mellor (1983)	Juvenile chronic arthritis	75 children	7-17	Consecutive attenders at juvenile rheumatology clinics at two hospitals.	Open-ended questions about illness beliefs during home interviews. Understanding categorized according to complexity of children's explanations. Drawings of the effect of JCA on their bodies. Fixed choice question about beliefs re. the value of treatment. No reliability or validity data.

Continued on next page

Table 3.1 continued

REFERENCE	SAMPLE CHARACTERISTICS			SAMPLING METHODS	MEASURING INSTRUMENT
	ILLNESS	SIZE	AGE OF CHILDREN		
Allen, Affleck, Tennen, McGrade and Ratzan (1984)	Juvenile diabetes	34 children	8-17	Attenders at a university hospital diabetic clinic.	Open-ended questions posed during a semi-structured interview. Responses coded by trained raters. Inter-rater reliability assessed between 0.72-0.92 using Kappa -coefficients.
Jamison, Lewis and Burish (1986)	Cancer	31 children with cancer	12-18	Attenders at a university hospital cancer clinic. Matched controls were drawn from local junior and senior high schools.	28-item knowledge of cancer scale. No evidence of reliability and validity although this scale has been widely used in previous studies.
Nolan, Desmond, Herlich and Hardy (1986)	Cystic fibrosis	28 patients 25 parents	10-21	Attenders at a specialist CF out-patient clinic.	22-item questionnaire. Content validity was provided by a representative panel of experts. No reliability data.

Continued on next page

Table 3.1 continued

REFERENCE	SAMPLE CHARACTERISTICS			SAMPLING METHODS	MEASURING INSTRUMENT
	ILLNESS	SIZE	AGE OF CHILDREN		
Spykerboer, Donnelly and Thong (1986)	Asthma	128 parents 110 controls	1-31	Randomly selected from children admitted to hospital with a definite diagnosis of asthma. Controls recruited from children who were hospitalized for minor surgical complaints.	77-item questionnaire on knowledge and misconceptions concerning asthma. No psychometric data. Administered by trained interviewers.
Harbord, Cross, Botica and Martin (1987)	Cystic Fibrosis	61 children	4-18	Attenders at a CF clinic.	Questionnaire on understanding of CF & its treatment. Administered by interviewers. Responses scored blind by 2 judges. No psychometric data.

their sample characteristics (chronic illness, size and age range), methods of sampling and measures of knowledge. The references appearing in Table 3.1 have been chosen because they include at least some operationalization and quantification (however simple and dubious) of the concept of knowledge. (Table 3.1 does not include those studies whose only purpose in measuring knowledge was to assess its association with compliance. These studies are reviewed in Section 3.2.3).

A number of studies have found that chronically ill children and young adults have unsatisfactory levels of knowledge about their conditions (Etzwiler 1962; Ludvigsson 1977; Martin, Landau and Phelan 1978; Reddihough, Landau, Jones and Rickards 1978; Johnson, Pollak, Silverstein et al 1982; Beales, Holt, Keen and Mellor 1983 and Lorenz, Christensen and Pichert 1985). Similarly low levels of knowledge have been identified amongst the parents of chronically ill children (Maxwell and Gane 1962; Kennell, Soraker, Thomas and Wasman 1968; Abdurrahman 1978; Reddihough, Landau, Jones and Rickards 1978; Conway and Littlewood 1985; Jackson, Glasgow and Thom 1985 and Spykerboer, Donnelly and Thong 1986). Parental knowledge of genetics was particularly inadequate (Taylor and Merrill 1970; Sibinga and Friedman 1971; Leonard, Chase and Childs 1972; Reiss and Menashe 1972; McCrae, Cull, Burton and Dodge 1973; Burton 1975 and McLucas, Hsia and Mick 1975).

3.2.2.1 Correlates of Knowledge in Chronically Ill Children

Evidence is categorical that AGE is a particularly important determinant of the chronically ill child's understanding of his illness (Etzwiler 1962; Partridge, Garner, Thompson and Cherry 1972; Ferencz, Wiegmann and Dunning 1980; Hamburg and Inoff 1982; Johnson, Pollak, Silverstein et al 1982; Susman, Hersh, Nannis et al 1982 and Nolan, Desmond, Herlich and Hardy 1986). One exception is noted in a pilot study of 12 haemophiliac families (Klein and Nimorwicz 1982) where it was found that children under 15 were better informed than children over 15 years. The small sample size, however, makes these findings questionable.

Various studies have highlighted the role of COGNITIVE DEVELOPMENT in the chronically ill child's understanding of the complex nature of his illness (Susman, Hersh, Nannis et al 1982; Allen, Affleck, Tennan et al 1984 and Harbord, Cross, Botica and Martin 1987), his understanding of the rationale for treatment (Beales, Holt, Keen and Mellor 1983 and Harbord, Cross, Botica and Martin 1987) and his acquisition of self-management skills (Johnson, Pollak, Silverstein et al 1982; Brown 1985 and Ingersoll, Orr, Herrold and Golden 1986). The findings that age and cognitive development are correlated with specific knowledge of chronically ill children is in keeping with the findings reviewed earlier concerning their knowledge of general illness and treatment concepts.

In 3 studies (Hamburg and Inoff 1982; Johnson, Pollak, Silverstein et al 1982 and Nolan, Desmond, Herlich and Hardy 1986) the SEX of the child has been found to predict levels of knowledge, with girls scoring consistently higher than boys. Johnson, Pollak, Silverstein et al (1982) suggest that this finding may be 'due to the greater developmental maturity that girls often demonstrate' (p.713). Alternatively, they suggest that girls tend to identify and spend more time with their mothers who, in turn, are more knowledgeable than fathers. These interpretations, however, remain speculative.

Whereas EDUCATION correlated positively with knowledge in two studies (Ferencz, Wiegmann and Dunning 1980 and Nolan, Desmond, Herlich and Hardy 1986), no relationship was found by Collier and Etzwiler 1971.

A positive relationship between PARENT'S KNOWLEDGE and that of their children was noted in two studies (Ludvigsson 1977 and Susman, Hersh, Nannis et al 1982). Patients knew significantly more than their parents in two samples (Partridge, Garner, Thompson and Cherry 1972 and Susman, Hersh, Nannis et al 1982). Susman and co-workers (1982) attributed this finding to the fact that their sample of cancer patients concentrated on the medical facts and technical procedures in their responses, whereas parents focused their responses on what might happen to the patient in the future.

DURATION of illness has not been found to predict levels of knowledge (Collier and Etzwiler 1971 and Johnson, Pollak, Silverstein et al 1982). Ludvigsson (1977) found a positive correlation between knowledge and POSITIVE ATTITUDES towards diabetes.

More recently research has begun to investigate the implications of knowledge for the chronically ill child's emotional well-being. Allen, Affleck, Tennen et al (1984) have noted that together with a more advanced understanding of his disease, the older child evidences correspondingly higher levels of concern about the future. These authors report a moderately high correlation ($r = 0.46$, $p = 0.002$) between more advanced disease concepts and worries about future complications in their sample of diabetic children. They conclude that while, on the one hand, the child is developing a more mature understanding of his disease which, in turn, leads to the acquisition of greater self-management skills, on the more painful side, these children also experience more disease-related anxieties. They do not, however, interpret these findings as evidence of psychopathology, but rather as a reflection of realistic fears considering the chronicity of their illness.

Jamison, Lewis and Burish (1986) also found that knowledge tends to be a double-edged sword. They noted that while knowledge of cancer was found to increase perceptions of cancer severity, it also brought a greater awareness of the benefits of treatment. Similar findings were reported by Beales, Holt, Keen and Mellor (1983). These researchers found that the older, cognitively more mature group (12-17 years) of children with juvenile chronic arthritis, reported significantly higher levels of distress (measured by visual analogue scales) about what was taking place inside their joints than the younger group (7-11 years). The former group, however, had a far greater appreciation of the rationale for their treatment.

On the basis of projective test findings, Tropauer, Franz and Dilgard (1970) report that the primary anxieties and conflicts related to CF, varied according to age and cognitive development. Using standardized measures of anxiety and depression, Klein and Nimorwicz (1982) found a negative relationship between knowledge of haemophilia and self-reported psychological distress in children and parents. Unfortunately the sample size (n = 12) was too small for generalization. Susman, Hersh, Nannis et al (1982) also found that hospitalized cancer patients and their parents were able to maintain an optimistic view of the future despite their knowledge that they might not have a future. A positive correlation between knowledge of cancer and self-image was reported by Jamison, Lewis and Burish (1986).

Having divided their sample of 35 CF patients (aged 16 to 51 years) into 'copers' and 'non-copers', Pinkerton, Trauer, Duncan et al (1985) found that 'copers' were better informed about their illness than 'non-copers'. Unfortunately on the basis of the reported findings, it is not possible to establish how 'coping' or 'knowledge of CF' were either defined or measured.

More recently research has begun to focus on the relationship between knowledge and compliance in chronically ill children and their parents. Because of the theoretical and clinical relevance of these studies, they will be reviewed separately later in the section.

In summary, a great deal is expected of the chronically ill child in terms of understanding and managing his illness. Research has shown that the chronically ill child's understanding of his illness and its treatment advances with his age and cognitive maturity. Educational level, sex and parent's knowledge have also consistently predicted levels of knowledge. Findings regarding the impact of improved understanding on the chronically ill child's emotional adjustment are scarce and equivocal. While some studies show that increased understanding leads to increased concern about the future, others have found a negative relationship between knowledge and psychological distress. Further research is needed to clarify the role of knowledge in the chronically ill child's adjustment to his illness.

3.2.2.2 Correlates of Parental Knowledge of Chronic Illness

Parental STATUS is a consistent predictor of knowledge, with mothers evidencing higher levels of knowledge than fathers (Etzwiler 1967; Burton 1975; Johnson, Pollak, Silverstein et al 1982; Klein and Nimorwicz 1982 and Nolan, Desmond, Herlich and Hardy 1986). This finding is generally ascribed to the fact that mothers tend to take their chronically ill children to clinic more frequently than fathers and therefore have greater opportunity to speak to the doctors (Burton 1975). Alternatively, because mothers are more involved in the day-to-day management of the chronically ill child, necessity may have prompted them to seek more answers to their problems. These interpretations, however, remain speculative.

Several studies report a positive correlation between SOCIAL CLASS and parental knowledge (Burton 1975; Jackson, Glasgow and Thom 1985 and Nolan, Desmond, Herlich and Hardy 1986), while McLucas, Hsia and Mick (1975) found no correlation. These discrepancies may well be attributable to differing measures of social class.

With the exception of Leonard, Chase and Childs (1972), other studies have not found a significant correlation between parental knowledge and EDUCATION (Collier and Etzwiler 1971; Sibinga and Friedman 1971; McLucas, Hsia and Mick 1975; Abdurrahman 1978 and Klein and Nimorwicz 1982). Sibinga and Friedman (1971) suggest that in view of the poor relationship between parental educational level and knowledge, the ability to understand a disease is more an emotional than a cognitive phenomenon. They propose that 'the degree of emotional resistance to the assimilation of medical information might be directly related to whether an illness is hereditary in nature ... whether the disease is culturally undesirable in outcome (e.g. mental retardation) ... or whether acknowledgement of the reality of the illness forces drastic changes in preplanned parental functioning' (p. 223). Accordingly different chronic illnesses will lead to different levels of parental understanding and adjustment.

Burton (1975) noted a positive association between parental understanding and their satisfaction with the explanations (about genetics) provided by the doctors.

No significant relationships between knowledge and the following variables have been reported: religion (Leonard, Chase and Childs 1972 and McLucas, Hsia and Mick 1975); duration of illness and the number of clinic attendances and hospitalizations (Abdurrahman 1978).

✳ In summary, only parental status (i.e. mother or father) and social class seem to predict parental knowledge, with the remaining variables showing no relationship whatsoever.

A word of caution is needed when interpreting the correlational evidence pertaining to patient and parent knowledge. The studies under review reveal complex interactions between variables such as interalia education level, social class and membership of parent associations. For example, Burton (1975) found that mothers of CF children had a more complete understanding of genetics than fathers. However, fathers in social classes I and II in general had the best understanding. It is possible that whatever correlations do emerge could be the result of co-variation between variables resulting in the confounding of true relationships. Many of these problems are best dealt with through the use of multivariate analysis.

It is also necessary, at this stage, to highlight the major methodological shortcomings of many of these studies.

3.2.2.3 Methodological Shortcomings of Empirical Studies of Patient and Parent Knowledge of Chronic Illness

It is a truism that in research the quality of the findings and the validity of any subsequent interpretations, are no better than the design and measurement methods employed.

Frequently occurring shortcomings include:

i. The lack of reliable and valid measures of knowledge. Only a handful of studies provide even a minimum of psychometric data concerning their measuring instruments. The most commonly reported type of validity, i.e. content validity, has been heavily criticised (Guion 1978 and Messick 1980). These authors point out that content validity is concerned more with test construction than with test performance. They feel that further evidence is required to interpret the meaning of test <u>scores</u>, for which construct validity is a prerequisite. Until studies use objective and psychometrically sound measuring instruments, replication and generalization of findings will remain a problem.

ii. Few studies provide definitions of their concept of knowledge and still fewer investigate knowledge within any sort of theoretical framework. This, in turn, renders the interpretation of findings very difficult.

iii. Measures rarely consider the complex, multidimensional nature of disease-related knowledge, the analysis of which requires an understanding of its many domains (Johnson 1984). For example, Johnson, Pollak, Silverstein et al (1982) found that knowledge of one area of diabetes did not necessarily predict knowledge of another aspect. Thus global, non-specific measures of knowledge are not adequate.

iv. In the majority of studies, subjects are drawn from convenience (e.g. specialist clinics) rather than random samples (Achenbach 1978).

v. Control groups are included in only two studies reported in Table 3.1.

Although valid, these criticisms must be viewed against the realistic difficulties encountered in chronic illness research. These typically include problems in validating instruments in samples of chronically ill patients which are often limited in size (Patterson 1984), in locating sizable samples of chronically ill children whose only common denominator is their clinical status (Masters, Cerreto and Mendlowitz 1983) and in using control or contrast groups from different chronic illnesses whose experiences are inevitably very different (CF Psychosocial GAP Conference Report 1981).

Future researchers will, nevertheless, have to concentrate their efforts on developing highly specific, psychometrically sound and theoretically justifiable measures of knowledge. Whenever possible these measures should be standardized within disease entities so that findings may be compared.

3.2.2.4 Knowledge of Cystic Fibrosis among Patients and Parents

For the sake of completeness, several studies which base their findings on 'impressionistic' assessments have also been included in this discussion.

Based on their findings from psychiatric and social work interviews with 20 CF families (including 26 CF children aged 10 years and under) Kulczycki, Robinson and Berg (1969) found that most parents had 'an intellectual understanding' of symptomatology, treatment procedures, genetics and prognosis. They described the children's understanding as ranging 'from a comprehensive, accurate view of CF to some concepts which were frankly bizarre' (p.324). Many children also said that they had CF because they had been bad.

Also using psychiatric interviews, Tropauer, Franz and Dilgard (1970) found that 15 out of 18 CF children had 'a reasonably good understanding of the nature of cystic fibrosis relative to the degree of intellectual maturity' (p.426). Most children realized that the disease was incurable which was in keeping with earlier findings of Lawler, Nakielny and Wright (1966). Occasional distortions were, however, noted in the younger children. For example, one 9-year old girl believed that 'fatness' in food infected her mucus, while another 10 year old boy described his illness as a disease that 'gets your lungs like termites in wood' (p.426). These distortions may well be accounted for on the basis of immature cognitive development, although the more likely explanation is one of inadequate information as many mothers reported finding it difficult to discuss the disease with their youngsters.

In 1976 Boyle and her co-workers, using a combination of interviews and projective tests, encountered strong resistance to any discussion of CF within their sample of 27 CF patients aged between 13 and 30 years. Approximately one third of the adolescent group claimed to know only the name of their illness, despite having had the disease explained to them by their doctors. Projective test findings, however, provided evidence of well-developed disease concepts. Nor did these adolescents want any further information about CF because 'you can't do anything about it and it is'nt going to change my way of living' (p.323). Indeed one patient who denied that he knew anything about his illness, went on to accurately portray all the organs, in proper relationship, involved in CF. When confronted with this, he countered that he had actually looked up CF in an encyclopaedia. These authors felt that their sample could benefit from more open communication both within the doctor-patient relationship and within the family. They also recommended group sessions which could offer 'practical and concrete' information on how to cope with daily concerns.

More recent studies by Bywater (1981) and Strauss and Wellisch (1981) also found adequate levels of CF knowledge in their samples aged 12 to 16 years and 18 to 33 years respectively. Neither study, however, indicated how or what knowledge was measured.

In a Canadian study Nolan, Desmond, Herlich and Hardy (1986) measured the knowledge of 28 CF patients and 25 parents. Using a standardized questionnaire, they assessed three areas of knowledge: pathophysiology, treatment and genetics and reproduction. Whereas knowledge of the first two sections was generally comprehensive and detailed, knowledge of reproductive risks and male sterility was less than adequate.

The most recent study (Harbord, Cross, Botica and Martin 1987) assessed the understanding of 61 Australian CF children from as young as 4 years old. All respondents in the oldest age group (12 - 18 years) knew that they had been born with the disease, almost 80% were aware that both the respiratory and digestive systems were implicated in the illness and approximately two-thirds understood the rationale for administering pancreatic enzyme supplementation, physiotherapy and antibiotics. Slightly less than half of this group, however, were aware of the function of bronchodilators. In contrast, only a minority of the youngest patients (4 - 6 years) correctly answered questions about organ involvement and treatment. The knowledge of children aged between 7 and 11 years fell mid-way between that of the youngest and oldest patients.

Most CF studies report uniformly low levels of genetic knowledge amongst parents and patients. In a sample of 76 families with various genetic conditions of whom 39 had at least one CF child, Leonard, Chase and Childs (1972) found that 25% of CF families had flawed understanding, while a further 25% had no understanding. The poorest levels of knowledge in the sample were recorded amongst the CF families who often had the knowledge (i.e. could answer the questions correctly) but without understanding.

In their sample of Irish and Scottish CF families (n = 100) McCrae, Cull, Burton and Dodge (1973) found that approximately 80% did not have an adequate understanding of the disease's genetic basis. Reporting anecdotally Allan, Townley and Phelan (1974) state that few CF mothers understood the genetics in relation to their unaffected children and only 8% had sought further information for their 16 children of marriageable age. Burton (1975), too, found low levels of comprehension amongst her sample of 53 CF families - only 20% had a complete understanding with another 20% claiming no knowledge of genetics at all.

Five items were devoted to assessing the genetic knowledge of parents and CF patients in the study by Nolan and colleagues (1986). These included questions on carrier detection, carrier status and recurrence risks. Older CF children (13 years and over) scored significantly higher than younger CF children (10-12 years) on genetics/reproduction items. Only one significant, though anticipated, difference was detected between the patient and parent scores on genetics items - significantly more parents knew that both parents had to carry the CF gene. Although not statistically significant, patients surprisingly obtained proportionately higher scores on the 3 items relating to risks of recurrence.

It is noteworthy that similarly low levels of comprehension of genetics have been reported amongst the parents of other chronic illness groups: congenital heart disease (Reiss and Menashe

1972), Duchenne muscular dystrophy (Taylor and Merrill 1970 and McLucas, Hsia and Mick 1975) and phenylketonuria (Sibinga and Friedman 1971).

In summary, the clinical and research evidence shows that CF patients and their families are reasonably well-informed in most areas of CF knowledge, with the exception of genetics and reproduction, where levels of comprehension are uniformly low. These findings must, however, be treated cautiously as only one study, namely that of Nolan and co-workers (1986), used a measuring instrument that could be replicated with any degree of confidence, albeit within a small sample.

3.2.3 Knowledge and Compliance

In many chronic and life-threatening illnesses, the quality and duration of survivorship may well depend on lifelong adherence to a demanding treatment regimen (Koocher 1984). Cystic fibrosis is no exception and the complex and time-consuming nature of the CF therapeutic regimen is well-described in the literature (Stern 1986). Recently research has begun to scientifically examine the contribution which knowledge of an illness and treatment regimen, makes to overall compliance with recommended therapies (Johnson 1984). Accordingly, the following section selectively reviews the paediatric literature which has examined this relationship. It also briefly touches on issues pertaining to inadvertent non-compliance, the causes of which can often be traced back to inadequate knowledge. The potential contribution which patient education makes to increasing knowledge and improving health outcomes is briefly assessed.

Because the relationship between knowledge and compliance has not been empirically explored in the CF literature, the following review has once again drawn on equivalent findings from other chronic illness groups. Where available, anecdotal findings from the CF literature are included in the discussion.

Haynes (1976) described compliance as 'one of the least understood yet most guessed about topics in health care' (p.5). This statement seems as true when evaluating the contribution which knowledge makes to compliance. Intuitively it seems that knowledge of a disease is a prerequisite for managing the disease; after all, if a patient or parent does not know what to do, they cannot do it (Johnson 1984). Studies have, however, failed to show conclusively that knowledge about an illness and its treatment automatically enhances compliance. Findings, based on predominantly adult samples, have varied from a positive relationship to no relationship at all (reviews by Haynes 1976; Masur 1981 and Becker 1985).

Haynes (1979) defines compliance 'as the extent to which a person's behaviour (in terms of taking medication, following diets, or executing lifestyle changes) coincides with medical or health advice' (p.1). Insofar as the literature on compliance in chronic paediatric illness has been thoroughly reviewed (Slope 1981 and Friedman and Litt 1986, 1987) and the conceptual and methodological problems in measuring patient compliance have also been summarized (Gordis 1979), the present discussion will limit itself to examining empirical studies which have specifically explored the relationship between knowledge and compliance in chronic paediatric illness.

A chronological summary of these studies is presented in Table 3.2, according to their sample characteristics, measures of knowledge and compliance, and selected findings. Once again, only studies which offer some operationalization and quantification of the concepts of knowledge and compliance are included in this review. The research reviewed is characterized by a wide spectrum of disease and a complete lack of consistency within the various measures of knowledge and compliance, making any comparison between studies very difficult.

Five (Christensen 1983; Smith, Seale and Shaw 1984; Conway and Littlewood 1985; Jackson, Glasgow and Thorn 1985 and McCowen, Hackett, Court and Parkin 1986) out of the ten studies show a positive link between knowledge and compliance. Certain methodological inadequacies are, however, present in these studies. In two of the samples (Smith, Seale and Shaw 1984 and Conway and Littlewood 1985) it is not possible to establish exactly what aspects of knowledge are being measured. On the basis of her doctoral dissertation findings, Slope (1981) concluded that only knowledge that is highly specific about what parents need to know in order to comply with treatment, is related to compliance. She did not find that more general knowledge concerning the illness, would promote compliance.

TABLE 3.2

KNOWLEDGE AND COMPLIANCE IN CHRONIC PAEDIATRIC ILLNESS : A SYNOPSIS OF RESEARCH STUDIES

REFERENCE	DESCRIPTION OF SAMPLE	MEASUREMENT OF KNOWLEDGE	MEASUREMENT OF COMPLIANCE	ASSOCIATION BETWEEN KNOWLEDGE AND COMPLIANCE
Gordis, Markowitz and Lilienfeld (1969)	136 children with a past history of rheumatic fever (9 - 13 years).	Items on rheumatic fever and penicillin therapy were included in a larger structured questionnaire. No psychometric data. Administered to parents.	Parental report.	No association.
Christensen (1983)	157 type I diabetic children (7-16 years).	Standardized knowledge of diabetes test administered to patients.	Self-management was assessed by a 10 item assessment form, by trained medical staff. Self-management referred to ability to manage urine and blood tests, interpret results, perform own injections, follow diet plan and recognize hypoglycaemic symptoms. The content of the assessment was validated by a panel of experts.	A significant association between knowledge and self management ($p < .05$).

Continued on next page

Table 3.2 continued

REFERENCE	DESCRIPTION OF SAMPLE	MEASUREMENT OF KNOWLEDGE	MEASUREMENT OF COMPLIANCE	ASSOCIATION BETWEEN KNOWLEDGE AND COMPLIANCE
Smith, Seale and Shaw (1984)	200 asthmatic children (1.5 - 17.3 years).	Subjective rating of parental comprehension of the illness and medication. No psychometric data.	Numerical index of compliance based on number of doses of prescribed medication. Validation data given.	General comprehension of illness significantly correlated with drug therapy ($p < 0.005$).
Conway and Littlewood (1985)	110 children who were hospitalized for acute asthma (0.8 - 14.4 years).	Parental understanding of asthmatics was rated as good, moderate or poor by a doctor. No details of content of knowledge.	Doctor rated compliance as good, inadequate, poor. No further details given.	12% of children whose parents were assessed as having poor understanding, were receiving adequate treatment at home, compared to 71% of children whose parental understanding was moderate to good.
Jackson, Glasgow and Thorn (1985)	50 children with coeliac disease (1.5 - 19 years).	8 item knowledge test administered to parents - included general questions and dietary knowledge. No psychometric data.	A self-report measure of compliance with diet - 3 options presented: strictly maintained, broken regularly or sometimes, diet kept poorly or ignored.	Poor dietary compliance was closely linked to poor parental knowledge ($p < 0.001$).

Continued on next page

Table 3.2 continued

REFERENCE	DESCRIPTION OF SAMPLE	MEASUREMENT OF KNOWLEDGE	MEASUREMENT OF COMPLIANCE	ASSOCIATION BETWEEN KNOWLEDGE AND COMPLIANCE
Lorenz, Christensen and Pichert (1985)	90 insulin-dependent diabetics (9 - 15 years).	i. Recall of patient's personal diet ii. Correctly filling a plate of food from a dietary sheet iii. Ability to choose a menu in a restuarant.	Error rates based on observation of actual eating behaviour. Ratings by trained medical staff.	No association.
Friedman, Litt, King et al (1986)	25 epileptic children (9-17 years).	3 item test of knowledge of treatment regimen - name of medications, number of pills per dose and number of daily doses. Administered to patients. No psychometric data.	Saliva phenobarbital samples. This method has proven reliability. Construct validity was established by showing significant correlations with 6 behavioural indications of compliance.	No significant correlation between saliva phenobarbital scores and knowledge of drug regimen (r = 0.02).
Jamison, Lewis and Burish (1986)	27 cancer patients (12-18 years).	28 item test of general cancer knowledge. No psychometric data. Administered to patients.	2 oncology nurses rated co-operation on a 10-item scale. Cronbach alpha 0.87 for both raters.	No significant correlation when age controlled for.

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Table 3.2 continued

REFERENCE	DESCRIPTION OF SAMPLE	MEASUREMENT OF KNOWLEDGE	MEASUREMENT OF COMPLIANCE	ASSOCIATION BETWEEN KNOWLEDGE AND COMPLIANCE
McCowen, Hackett, Court and Parkin (1986)	96 diabetic children, 152 mothers, 62 fathers (11 years and older).	83 item multiple choice test of diabetes knowledge. No psychometric data.	Dietary compliance was assessed by means of 3 day diary kept by parents, patients or both. Compliance was assessed by comparing daily diet with the prescribed diet. Previous studies have proven the suitability of this measure.	Only mother's test scores were weakly correlated with aspects of the patients' dietary intake.
Wynn and Eckel (1986)	16 children with juvenile chronic arthritis (4-16 years).	Questions covering knowledge of home therapy were included in a broader questionnaire. No psychometric data.	Parental report in answer to question 'How often does your child follow the recommended program?' Compliance ratings were assigned numerical values of 1 to 5, for poor through complete compliance.	No association.

From these two studies it is not possible to relate specific knowledge to drug compliance. McCowen and co-workers (1986), however, found a direct relationship between mothers' knowledge of diet and dietary compliance. Both Jackson, Glasgow and Thorn (1985) and Conway and Littlewood (1985) had also relied on unvalidated self-report measures of compliance. Only one study reported a positive association between patient knowledge and self-management (Christensen 1983). The fact that the study was conducted at a diabetic camp may have biased these findings as self-management was actively encouraged by the staff. Patients may not necessarily practise self-management in other settings.

Certain trends did, nevertheless, emerge. In 4 out of the 5 studies with positive findings, it was parental and not patient knowledge that correlated with compliance, suggesting that parental knowledge may be a better predictor of compliance. Clearly the finding that mothers' dietary knowledge is linked with dietary compliance supports this suggestion, as mothers are characteristically responsible for preparing family meals, irrespective of a child's age. Furthermore, 3 samples (Smith et al 1984; Conway and Littlewood 1985 and Jackson et al 1985) with positive findings included much younger children, whose compliance would clearly be dependent on parental control. Unfortunately these studies did not explore the relationship between knowledge and compliance while controlling for patients' age and developmental level.

A further cause for concern amongst these studies arises from the co-variation between many of the measured variables such as, for example, social class and membership of a Parent Association. As was noted in the previous section, whatever relationships do emerge may simply result from spurious correlations between these variables. Unless the effects of intercorrelated variables are controlled for, knowledge-compliance linkages may be obscured.

The mixed findings from these studies preclude any definite conclusions being reached about the relationship between knowledge and compliance in chronically ill children.

3.2.3.1 Knowledge and Compliance in CF studies

No formal assessment of the relationship between knowledge and compliance has been undertaken among CF samples. While Nolan, Desmond, Herlich and Hardy (1986) measured knowledge of treatment amongst CF patients and their parents, they did not link their findings to a measure of compliance. Nonetheless, their findings do highlight certain inadequacies in CF patients' knowledge of treatment which might well have some bearing on compliance. For example, while 82% of patients knew why they needed aerosol therapy, a mere 14% correctly answered a similar question concerning vitamin supplementation. Furthermore, although 68% of patients knew why they were taking enzyme supplements, only 46% knew what would happen if they stopped taking these supplements - this being a clear example of knowledge without understanding.

Yet other CF studies provide anecdotal evidence both negating and supporting a link between knowledge and compliance. Based on their findings from psychiatric interviews, Tropauer, Franz and Dilgard (1970) observed that an accurate and realistic concept of CF was no guarantee of compliance with treatment, as many difficulties with therapy were encountered with 'highly intelligent' youngsters (p.426).

Burton (1975) using a sample of Irish CF families, noted 'the considerable benefit in terms of the child's response to treatment which accrued from proper supportive parental explanations' (p.96). She found that in general, where the CF children were aware that treatment was being administered in their best interests, their protests seemed to be less vehement. In contrast 'lies, evasions ... on the part of the parents often produced near panic reactions in the child' (p.94) which occasionally led to treatment refusal. Furthermore, inadequate explanations were also linked to distorted thinking on the part of the child. Burton (1975) cites the example of a child who refused mist tent therapy because she thought the mist tent was a cellophane bag which would lead to suffocation. Burton (1975) also found that where explanations about treatment contained an element of fear, resistance to treatment increased. She gives the example of a mother who told the young patient that she had to have physiotherapy to remove 'a wee mouse down in your chest' (p.90). Far, be it from improving compliance, the child expressed more fears about her illness and complied less.

While these anecdotal accounts provide valuable insight into the possibility of an association between knowledge and compliance in CF samples, they are insufficient to reach any conclusions about the relationship between these two variables.

3.2.3.2 Suggestions for Future Research

Future research which examines the relationship between knowledge and compliance will need to use more psychometrically sound measures which are sensitive to the multidimensional nature of both compliance and knowledge. Masur (1981) points to the need for future research to elaborate on exactly what aspects of knowledge, compliance is being measured against. In much of the present research, 'knowledge' has been variously defined as meaning specific information about a treatment regimen, general medical information or simply knowledge about the rationale for treatment (Eraker, Kirsch and Becker 1984). Until it is clear what researchers mean by 'knowledge', it will be difficult to compare findings of studies which have attempted to evaluate its potential role in enhancing compliance.

Future studies will also need to specify the direction of the relationship between knowledge and compliance at the outset of the study. While a few studies have indicated a slight positive relationship between these two variables, it is equally conceivable that in a life-threatening illness such as CF, specific knowledge of, for example, prognosis or the questionable efficacy of certain treatment, may lead to lower levels of compliance - as Charney (1972) points out that the more fatalistically inclined patient may choose to 'tune-out' advice about treatment (p.273).

On the other hand, a strongly negative relationship between diabetes knowledge and metabolic control, led Hamburg and Inoff (1982) to suggest that poorly controlled patients sought to add to their knowledge of their illness in order to improve their medical status.

Thus only a priori hypothesizing will help to clarify how knowledge influences and is, in turn, influenced by health-related variables such as compliance.

3.2.3.3 Inadvertent and Voluntary Non-compliance

These two concepts add yet another dimension to the complex relationship between knowledge and compliance. 'Inadvertent noncompliance occurs when the patient believes he or she is adhering to recommended treatment but through errors in knowledge or skill, is actually noncompliant' (Johnson 1984) (p.514). Reasons for inadvertent non-compliance include poor understanding, the complexity of the treatment regimen or misconceptions about the illness and its treatment.

The true extent of inadvertent noncompliance in CF is unknown. However, high levels of self-medication with vitamins, antibiotics and non-prescribed drugs have been reported in 2 CF compliance studies (Meyers, Dolan and Mueller 1975 and Passero, Remor and Salomon 1981). These findings suggest a tendency on the part of parents and patients to overestimate the value of treatment. A similar finding was noted by Nolan and co-workers (1986) who found that 60% of patients thought they would die if they stopped treatment (antibiotics and physiotherapy) altogether.

The importance of an accurate knowledge for compliance is illustrated by the commonly held misconceptions among asthma patients and their parents (Martin, Landau and Phelan 1978 and Spykerboer, Donnelly and Thong 1986). Further, inaccurate knowledge can lead to medication errors (Martin, Landau and Phelan 1978; Ellis and Friend 1985 and James, Phillips and Thompson 1985), unnecessary restrictions on a patient's physical activity (Martin, Landau and Phelan 1978; Reddihough, Landau, Jones and Rickards 1978 and Spykerboer, Donnelly and Thong 1986) or even total disruption of family life. Spykerboer and colleagues (1986) observed that 54% of mothers erroneously believed that moving to the country would improve their children's asthma. Studies of diabetic children have found serious errors in urine testing, self-injection, blood glucose tests and choice of diet (Khurani 1971; Johnson, Pollak, Silverstein et al 1982; Lorenz, Christensen and Pichert 1985 and McCowen, Hackett, Court and Parkin 1986).

Whereas the inadvertent non-complier has the intent to comply with medical advice but because of misinformation or ignorance fails to do so, the voluntary non-complier has the necessary knowledge and skills but actively chooses not to follow the prescribed regimen (Conrad 1985). Explanations for voluntary non-compliance include the distress caused to the child by continuance of treatment (Beales, Holt, Keen and Mellor 1983 and Davids 1986) the repugnant nature of the treatment (De Wet 1984) and a desire to improve the child's quality of life (Deaton 1985).

Thus what might well appear to be a failure to comply from a medical perspective, may actually be an active decision not to comply based on the patient and parent's medical knowledge of the illness and their experience of the treatment. Deaton (1985) refers to this active weighing up of treatment options as 'adaptive non-compliance', which she feels is in sharp contrast to the more traditional perspective that views non-compliance as resulting from ignorance, forgetfulness, irresponsibility or carelessness.

In summary, these studies have shown that while being well-informed about an illness does not necessarily guarantee compliance, being misinformed may well lead to serious over-or-under medication. Clearly, knowledge is simply one of many variables that affect the patient or parent's decision and ability to comply.

3.2.4 Increasing Patient and Parent Knowledge about Chronic Illness

Given the increasing attention being focused on knowledge as a potential explanatory variable in a family's overall adjustment to chronic paediatric illness (Johnson 1984) an obvious next question would be whether it would benefit the patient and his parents to actively increase their levels of knowledge. Certainly the evidence reviewed so far has revealed inconclusive and mixed findings in measured relationships between knowledge and compliance and knowledge and emotional adjustment (i.e. anxiety and depression).

Nevertheless, patient and parent education has become an accepted component of chronic disease management (Friedman and Litt 1986). It aims ideally at teaching patients and parents not only to know something but also to do something (Nader 1985). Thus behaviour, such as compliance, becomes the objective of many educational efforts (Nader 1985). The rationale behind these educational efforts is the increase of disease-related knowledge and skills which should, in turn, translate into improved coping and self-management on the part of the patient (Wilson-Pessano and McNabb 1985).

Educational programmes reported in the literature have, however, produced mixed findings. While increased knowledge and compliance, as well as reduced morbidity, following an intensive programme of education, have been reported in some samples of asthmatic children (Fireman, Friday, Gina et al 1981; Richards, Church, Roberts et al 1981; Lewis, Rachelefsky, de la Sota and Kaplan 1984; McNabb, Wilson-Pessano, Hughes and Scamagas 1985 and Rubin, Leventhal, Sadock et al 1986), others have only reported increases in knowledge with no corresponding reduction in morbidity (Rakos, Grodek and Mack 1985; Hilton, Anderson, Sibbald and Freeling 1986 and Van Asperen, Jandera, De Neef et al 1986).

Similarly conflicting results are reported in the diabetic literature. Whereas most efforts at patient education led to increased levels of knowledge about diabetes and its management (Etzwiler and Robb 1972; Gilbert 1982 and Harkavy, Johnson, Silverstein et al 1983), this knowledge did not necessarily lead to increased dietary adherence (Lebovitz, Ellis and Skyler 1978) or improved control (Etzwiler and Robb 1972). Golden, Herrold and Orr (1986), however, managed to reduce the number of ketoacidotic episodes through a programme of intensive medical, educational and psychosocial intervention among patients and their families. Attempts at improving compliance by increasing knowledge of medication were not successful in a group of renal transplant patients (Beck, Fennell, Yost et al 1980).

While methodological shortcomings plague many of these studies (Popham and Yalom 1983 and Wilson-Pessano and McNabb 1985), it is clear that the most successful programmes are those which combine education and behavioural strategies (Mazzuca 1982). Wilson-Pessano and McNabb (1985) stress that patient education must go beyond teaching patients about their disease and treatment. They maintain that 'simply giving information may increase a patient's knowledge, but it may not bring about changes in health behavior required to remedy chronic management problems' (p.671). Similar sentiments were expressed by Mazzuca (1982) who said that 'patients need to know less about the pathophysiology of their disease and more about integrating new demands into their daily routine' (p.528).

In the only reported CF study on patient education in the published literature, Spirito, Russo and Masek (1984) were able to reduce anxiety and pain and improve sleep patterns in a sample of hospitalized CF patients, by teaching them selected behavioural and stress management skills. A complete and critical review of the large body of literature which has developed around the different techniques and methods of imparting knowledge and social/behavioural skills is, however, beyond the scope of this review.

Finally, these studies have shown that while patient and parent education generally leads to increased levels of knowledge, these educational efforts do not necessarily lead to improved health outcomes. To be successful these programmes must focus on improving both knowledge and social/behavioural skills. With few exceptions, patient and parent education has received scant attention in the published CF literature.

In summary, this review has focused on what sick and healthy children understand about medical matters in general and what chronically ill children and their parents understand about their particular illness. The major correlates of this knowledge were identified and the limited findings concerning the impact of knowledge on coping were evaluated. Because of its particular clinical significance, the relationship between knowledge and compliance was singled out for a more in-depth review. The potential role of patient and parent education in increasing knowledge and improving overall coping was briefly touched upon.

The remainder of this chapter focuses on the measurement and findings related to levels of CF knowledge in the present sample of CF families.

3.3 MEASUREMENT OF CF KNOWLEDGE, PERCEPTION OF PRESENT AND FUTURE HEALTH AND THEIR RELATIONSHIP TO SELECTED SOCIODEMOGRAPHIC AND MEDICAL VARIABLES

3.3.1 CF Knowledge Test

Following an unsuccessful search of the literature for a suitable, psychometrically sound measure of CF knowledge, a new instrument was developed for this purpose. An outline of the methodology employed in the construction of the CF Knowledge Test is presented in Table 3.3.

TABLE 3.3

OUTLINE OF STAGES OF DEVELOPMENT OF CF KNOWLEDGE TEST

1. Design Decisions
    - i. criterion - versus norm-referenced testing
    - ii. item format
2. Review of medical and lay literature to identify main content areas to be included in the test
3. Development of an initial pool of 120 test items
4. Preliminary construction of a 90 item Test
5. Internal review and modification by experts
6. Final draft of Test
7. Analysis of psychometric properties

As the purpose of the Test was primarily to identify how much families know about CF, the author designed a criterion-referenced test. The increased descriptive power of a criterion-referenced test makes it a more suitable measure than the traditional norm-referenced test in the health field (Popham 1982). According to Popham (1976) the criterion-referenced test can provide 'a precise fix on what an examinee can or cannot do' (p.593), whereas the primary aim of the norm-referenced test is to discriminate the good and the bad examinee. Testing should, furthermore, be two-directional, pointing to the possession of knowledge AND towards the diagnosis of gaps in this knowledge (Messick 1981). The criterion-referenced test is far better suited to this than the norm-referenced test.

A second crucial distinction between the criterion- and norm-referenced test centres on the extent to which tests must result in varied examinee performances. In the criterion-referenced test, items are chosen because they constitute a representative sample of the knowledge being tested, and not for their ability to discriminate between respondents. Thus low response variance is often a feature of the criterion-referenced test (Hambleton 1982). This latter characteristic was important in the present study as it was not possible, at the outset, to predict to what degree levels of knowledge would vary. Furthermore, the criterion-referenced test allows for the inclusion of relatively easy questions which might well be answered by over 90% of individuals. To have excluded items simply because of their low response variance might well have resulted in an unbalanced measure of CF knowledge (Basch and Gold 1985).

Although objective tests have been the subject of much research in the field of higher education, it is only recently that the psychometric properties of different item formats have received attention in the health care literature (Windsor, Roseman, Gartseff and Kerk 1981; Dunn, Bryson, Hoskins et al 1984 and Price, O'Connell and Kukulka 1985). These studies concluded that the multiple choice format was likely to reflect, most accurately, levels of knowledge. Although the true-false format appealed to those with poorer reading skills, it was more susceptible to guessing. All CF Test items were therefore designed according to the multiple choice format. Each item consisted of a main stem, a correct answer, one distractor and an 'I don't know' option. The purpose of the distractor was to discourage guessing.

Following an extensive review of medical and lay CF literature, an initial pool of 120 items was developed, from which 90 questions were selected for inclusion in a preliminary questionnaire. These were then submitted to three senior consultant paediatricians in the CF clinic to evaluate the accuracy of item content and to indicate, for each question, whether it was likely to be correctly answered by the parents at the CF clinic. (The final questionnaire included only two items, Nos 17 and 31, which the paediatricians felt might not be correctly answered by parents). The questions on genetics were also screened by a paediatric geneticist. To ensure the technical adequacy of the items, preliminary drafts of the CF Knowledge Test were evaluated, on two occasions, by an expert in the field of education. This resulted in the removal of badly worded and ambiguous items.

The final CF Knowledge Test contained 63 questions which covered the following content areas:

- general information about CF
- respiratory symptomatology and treatment (in CF)
- physiotherapy
- gastrointestinal symptomatology and treatment (in CF)
- nutrition
- genetics
- anatomy
- common CF terminology
- general medical information

The latter category included simple medical questions which were relevant to the daily management of a CF child. Almost 86% of the questions measured recall of knowledge, of which 16% also assessed commonly held misconceptions. The remaining 14% of items measured the application of CF knowledge (Appendix 7).

3.3.1.1 Psychometric Properties

a. Reliability

Reliability analysis (Kerlinger 1975) was performed on the composite CF Knowledge Test scores of parents, patients and siblings. Reliability coefficients (Cronbach Alpha) of 0.91, 0.94 and 0.95 respectively indicated a high level of internal consistency among the performances of all family groupings (Nelson 1980). Ebel (1979), furthermore, felt that since each cluster of items on a criterion-referenced test was most likely measuring different areas of knowledge, multiple reliability coefficients were necessary. Thus the reliability of the 9 sub-sections of the CF Knowledge Test was also assessed. (This analysis was performed on parental Test scores only). Reliability findings according to each sub-section were as follows: general information about CF (alpha 0.70), respiratory symptomatology and treatment (alpha 0.49), physiotherapy (alpha 0.54), GIT symptomatology and treatment (alpha 0.59), nutrition (alpha 0.75), genetics (alpha 0.66), anatomy (alpha 0.91), terminology (alpha 0.53) and general medical information (alpha 0.37). Given the small number of items in many of the sub-sections, lower reliability coefficients were not unexpected (Nelson 1980). With the exception of respiratory symptomatology (alpha 0.49) and general medical information (alpha 0.37), the remaining alpha levels were, however, above 0.50 which is the recommended minimum level if findings are to be used for between group comparisons (Helmstadter 1964).

b. Validity

Content validity refers to the representativeness of the questions included in the measuring instrument (Hambleton 1982). The content validity of the present study referred to the relationship between medical knowledge of CF and the Test content. Content validity was established by the process of test construction, during which items were sampled from an extensive search of professional and lay literature. The content of items was also evaluated by experts in the field of CF. Hambleton (1982) felt that content validity was further enhanced by the technical adequacy of the items. As already mentioned, the opinion of experts led to the removal of faulty items.

Construct validity refers to the ability of a measuring instrument to measure the specific construct it was designed to measure (Kerlinger 1975). It was, therefore, necessary to establish if the Test were indeed measuring knowledge of CF. Messick (1980) felt that a valuable source of construct validity could be provided by studying some differences among groups believed to differ in their levels of content mastery. Accordingly the CF Knowledge Test was administered to the following criterion groups who were thought to possess varying levels of CF knowledge:

18 paediatricians and senior paediatric registrars
8 physiotherapists
8 nursing sisters who were completing a one year diploma course in paediatrics
32 IVth year medical students who were completing a four week paediatric block
14 clerical and domestic staff at the Red Cross War Memorial Children's Hospital whose inclusion was determined by their professed lack of knowledge about CF.

A summary of the Test performances of the 5 criterion groups, together with equivalent findings from the study sample of CF mothers, is contained in Appendix 8. As was expected, none of the criterion groups which had received some training in medical matters obtained total mean scores (% correct answers) lower than mothers of CF children (Figure 3.1). In contrast, the non-medical respondents scored considerably lower than mothers.

Similar trends were observed when the performances of the paediatricians, physiotherapists, CF mothers and non-medical respondents were compared according to areas of content (Figure 3.2). Paediatricians scored predictably higher than CF mothers and non-medical respondents on all sub-sections of the Test. Chi-square analyses revealed that differences in performance between paediatricians and CF mothers were not significant on only two sub-sections, namely, general CF knowledge and physiotherapy. Differences in scores between paediatricians and the non-medical group were statistically significant on all sub-sections (p values for both sets of comparisons ranged between 0.01 and 0.0001). With the exception of the sub-section concerned with gastrointestinal symptoms and treatment, physiotherapists also scored consistently higher than mothers on all sub-sections. These differences were statistically significant in respect of nutrition ($p < 0.05$), anatomy ($p < 0.01$), terminology and general medical information ($p < 0.0001$). Physiotherapists scored significantly higher than the non-medical group on all sub-sections (p values ranged between 0.05 and 0.0001). (For the purpose of these Chi-square analyses, groups were compared according to number of correct versus incorrect/I don't know responses).

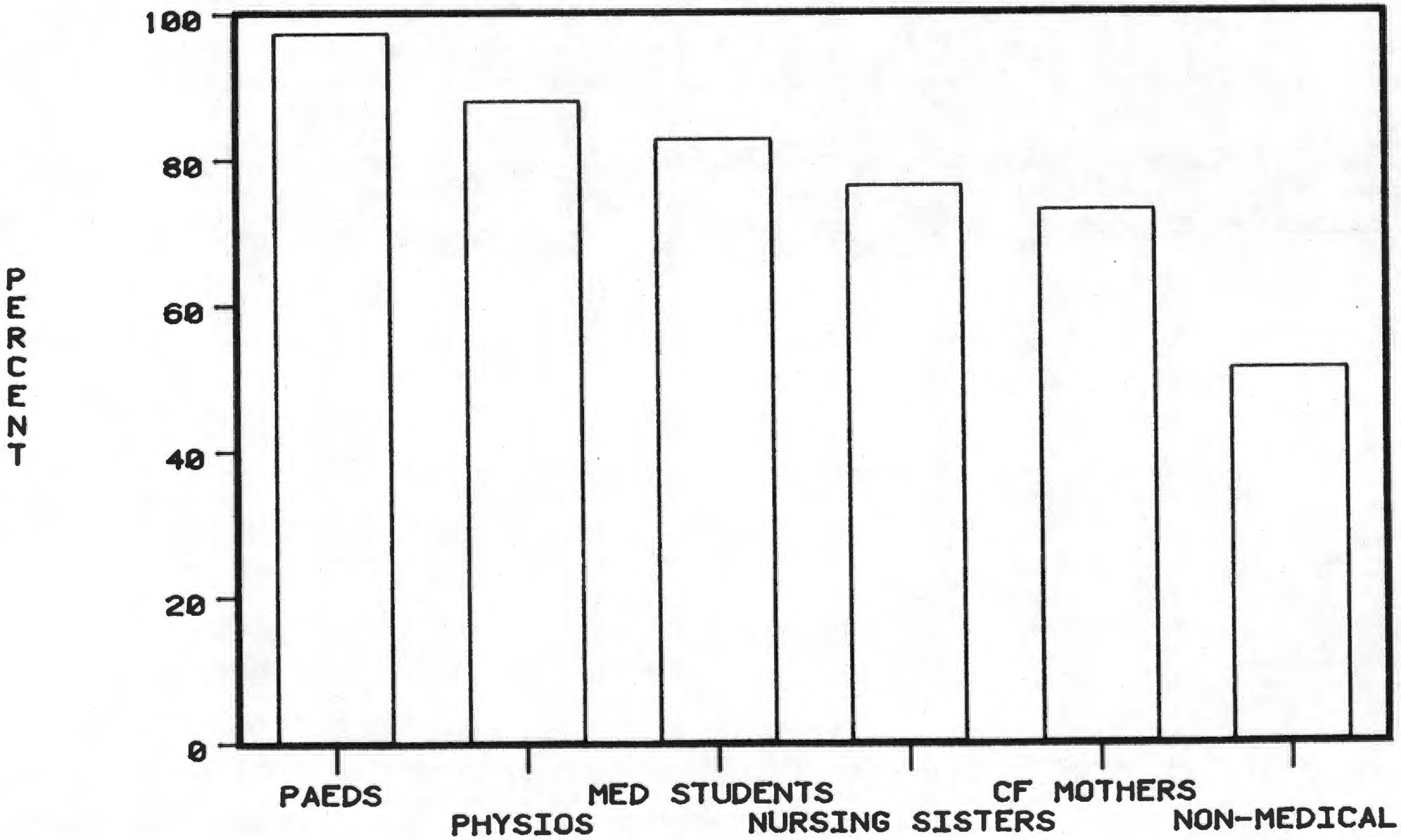

FIGURE 3.1: Total Mean Scores on the CF Knowledge Test (% Correct) for Paediatricians, Physiotherapists, IVth year Medical Students, Nursing Sisters, CF Mothers and Non-Medical Hospital Staff.

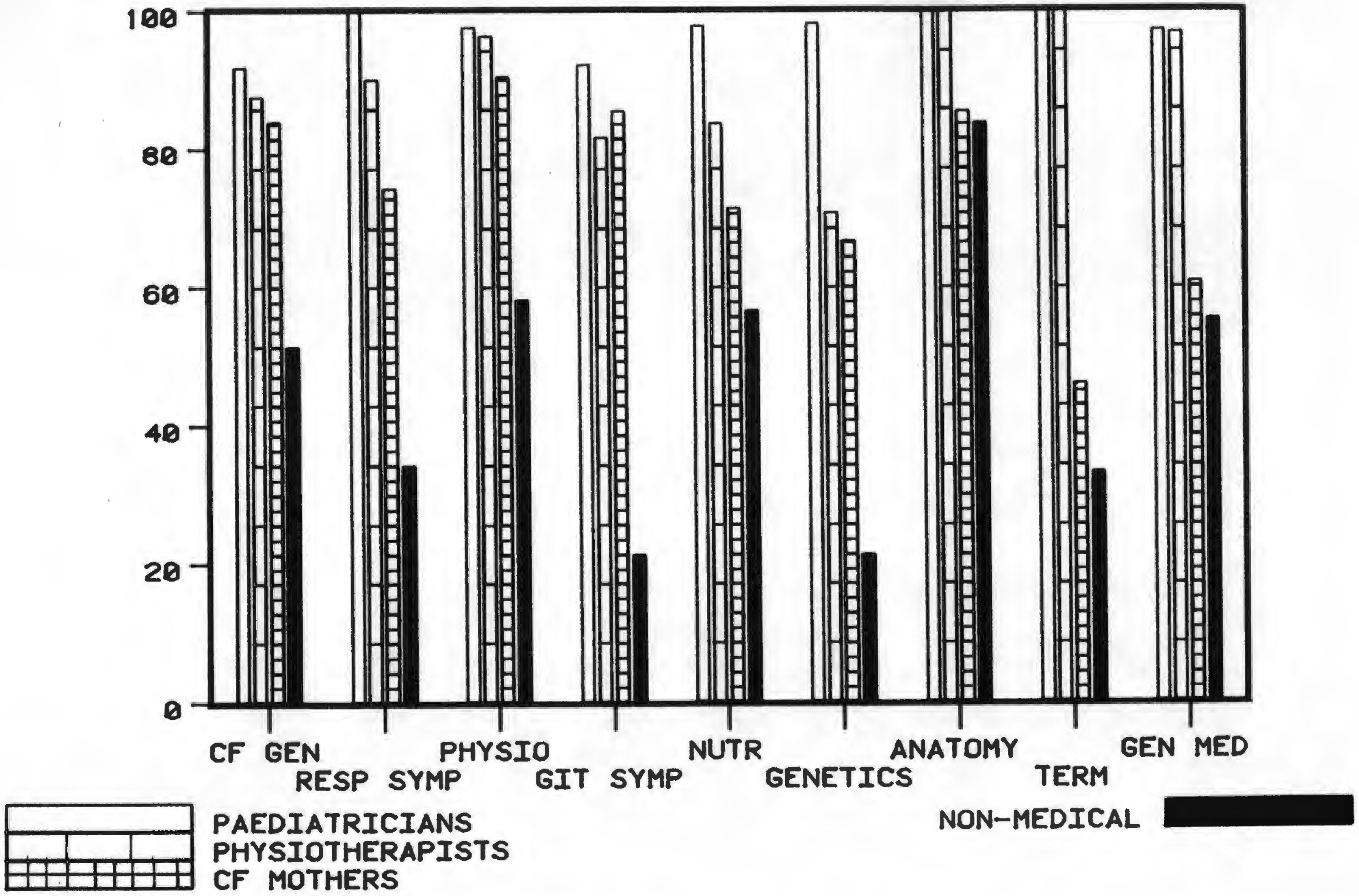

FIGURE 3.2: Mean Sub-section Scores on the CF Knowledge Test (% Correct) for Paediatricians, Physiotherapists, CF Mothers and Non-Medical Hospital Staff.

The CF Knowledge Test was also able to discriminate between persons with a specialized knowledge of CF and those with a more general medical knowledge. Paediatricians scored consistently higher than nursing sisters and IVth year medical students on the 6 sub-sections which related specifically to CF (Figure 3.3). Furthermore, it was important that the Test did not confound the constructs of general medical knowledge and specialized knowledge of CF. All criterion groups with some medical training received scores of one hundred per cent on sub-sections which measured general medical knowledge (Figure 3.4). With the exception of anatomy, mothers and non-medical respondents performed poorly on these sections. The Test was therefore able to detect differences in CF knowledge and general medical knowledge in this study sample.

Threats to validity: The validity of the Test findings, as true measures of CF knowledge, was threatened in the present study by the possibility of 'cheating' and guessing. Families were required to complete the questionnaires in the privacy of their homes which meant that there was no control over whether family members exchanged answers or looked up the correct answers in books or pamphlets. The need for family members to answer the medical items spontaneously and independently was addressed through the written Test instructions and again through verbal explanation when the questionnaires were delivered to the families.

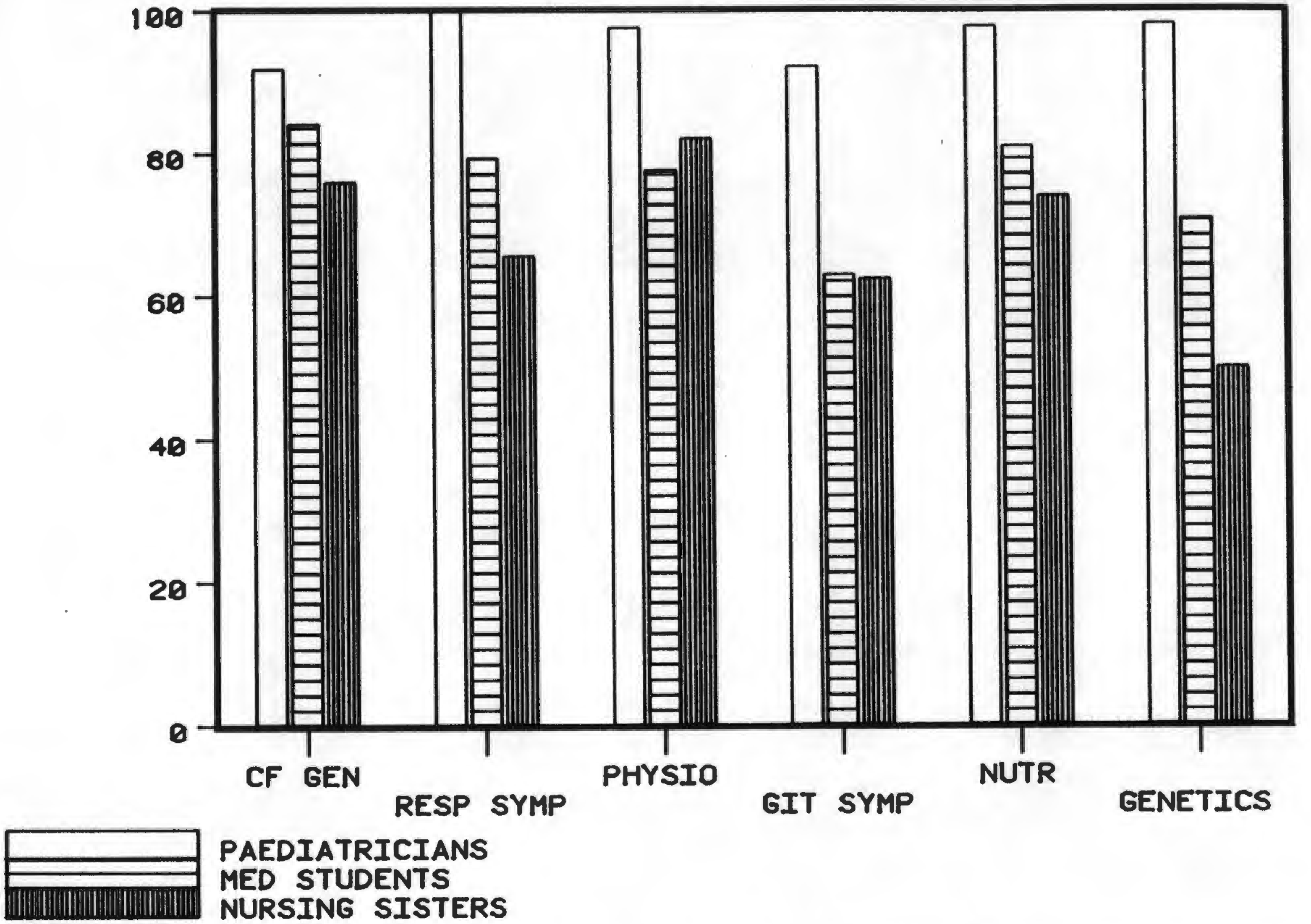

FIGURE 3.3: Mean Scores on the Sub-Sections Measuring Specialized CF Knowledge (% Correct) for Paediatricians, IVth year Medical Students and Nursing Sisters.

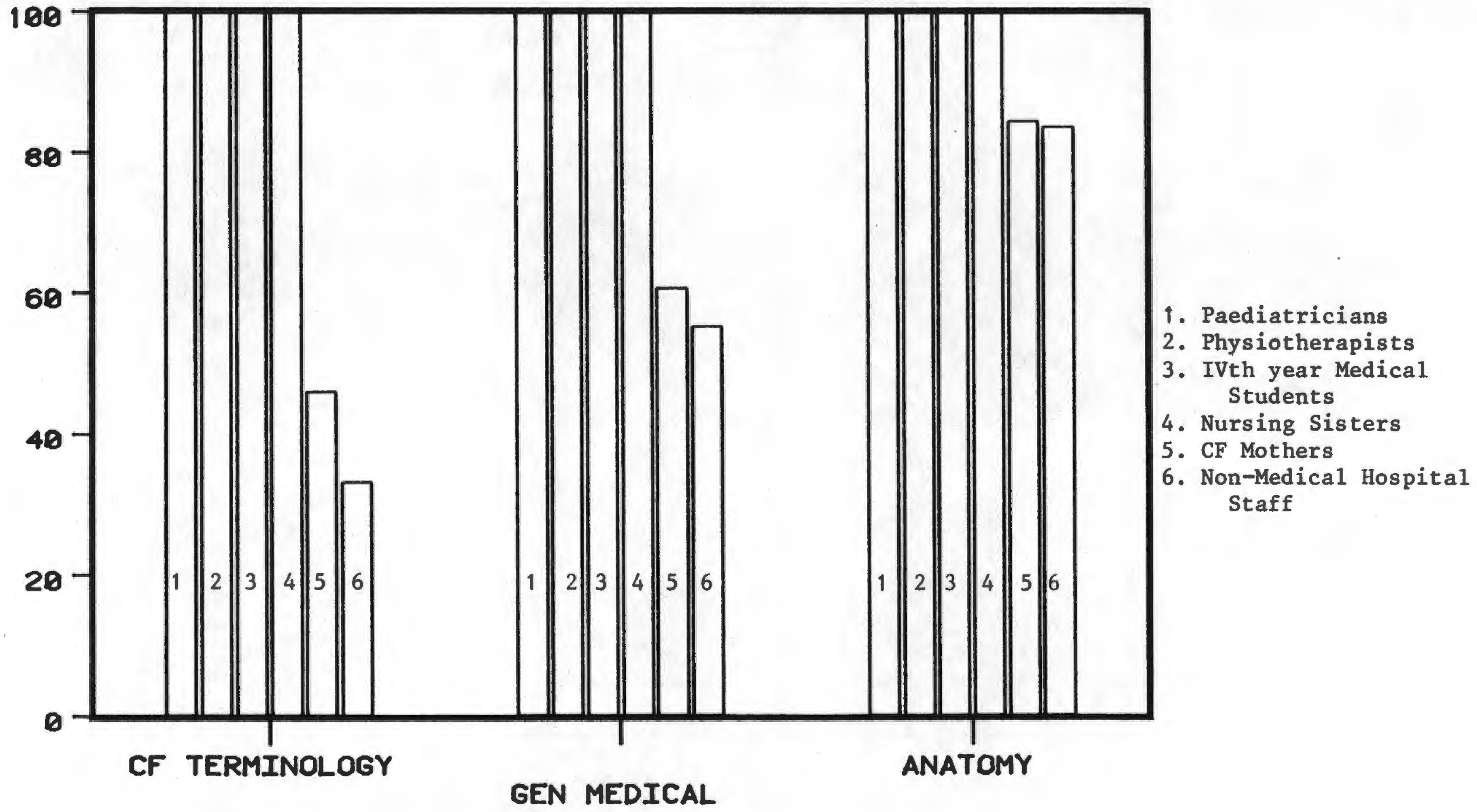

FIGURE 3.4: Mean Scores on the Sub-Sections Measuring General Medical Knowledge (% correct) for Paediatricians, Physiotherapists, IVth Year Medical Students, Nursing Sisters, CF Mothers and Non-Medical Hospital Staff.

Furthermore, it was hoped that by including the CF Knowledge Test within the comprehensive questionnaire, aspects of 'test anxiety' and possible feelings of inadequacy, caused by the evaluative component of the Test, would be reduced and thereby also reducing the tendency to guess instead of using the 'I don't know' option.

3.3.2 Choice of Sociodemographic and Medical Variables

The choice of variables was governed by the exploratory nature of the study and the need to confirm similar, though conflicting, findings from related research (see Literature Review). Accordingly the following sociodemographic and medical variables were considered in the study:

a. AGE (of the patient): As determined by his last birthday in round figures. Where a patient was 3 months from his next birthday, the age at the next birthday was recorded.

b. GENDER: Male or Female

c. RACE: Caucasian or Mixed

d. LANGUAGE: English or Afrikaans.

e. SOCIAL CLASS: As determined by father's occupation in accordance with guidelines outlined by the British Registrar General's Grading Scheme (Davie, Butler and Goldstein 1972) (pp 2-6). This classification ranges from Social Class I which includes professions requiring the highest qualifications to Social Class V consisting of unskilled manual labour. In a few exceptions where the mother was the breadwinner, her occupation determined the choice of social class and in the case of unemployed fathers, choice of social class was dependent on their most recent previous employment.

f. EDUCATION (of patients and siblings): As determined by number of completed years of education.

The reasons for excluding direct measures of parental education were two-fold. Firstly, to minimize test anxiety which could be raised by parents who might have erroneously associated their lack of knowledge on the CF Test with their poorer education. (Note: the education levels of patients and siblings were known to the author prior to the commencement of the study. This was not the case for parents). Secondly, social class (i.e. occupational level) has been shown to be moderately correlated with education and directly related to qualifications and training (Abramson, Gofin, Habib, Pridan and Gofin 1982).

g. MARITAL STATUS: Married or Single

h. SHWACHMAN SCORE: As determined by the method of Shwachman and Kulczycki (1958) which grades patients into excellent, good, mild, moderate and severe categories according to scores based on their general activity (e.g. school attendance, vigour of play, fatigue), physical findings (e.g. cough, clubbing), nutritional status (e.g. height vs weight, stool appearance) and chest X rays. A lower Shwachman Score indicates that a child is more severely affected by his illness. Scores for each patient were calculated by consultants in the CF clinic.

i. PERIOD SINCE DIAGNOSIS. Duration in months since a formal medical diagnosis was made according to the sweat test.

j. NUMBER OF DAYS IN HOSPITAL. Days spent in hospital between January 1, 1984 and June 30, 1986 as determined by hospital computer records.

k. NUMBER OF CLINIC ATTENDANCES: Between January 1, 1984 and June 30, 1986 as determined by hospital computer records.

3.3.3 Perception of Present and Future Health

Whereas the objective, multiple choice format of the CF Knowledge Test was an appropriate measure of family members' factual medical knowledge about CF, it was unsuitable to assess their more subjective knowledge of a patient's present and future health, the more so given the sensitive and painful nature of these perceptions. Accordingly two single item measures were constructed for this purpose. These ratings were adapted from a broad study of children's health status (Eisen, Ware, Donald and Brook 1979). These authors were of the opinion that general health ratings assessed both the objective information parents have about their child's health and their evaluation of that information.

3.3.3.1 Perception of Present Health

Family members had to indicate whether they considered the patient's present health to be excellent, very good, good, fair, poor or very poor.

3.2.3.2 Perception of Future Health

Family members were asked to rate, on a 7 point scale, how they viewed the patient's future health. Scale points were defined as cured, very much better, a little better, remain the same, a little worse, a lot worse, a great deal worse. For the purpose of the present study, this measure was also used as an indirect measure of perceived prognosis (Frydman 1981).

For both measures, high scores reflected more positive health assessments.

3.4 RESULTS

3.4.1 Description of Study Sample

The study sample comprised a total of 60 families, 6 (10%) of which were one parent units. (The response rate among families has already been discussed in Chapter 2). In 15 (25%) families the patient was an only child. Twenty eight (46.6%) families had 1 healthy sibling, 12 (20%) had 2 unaffected siblings while a further 5 (8.33%) had 3 unaffected children. The sociodemographic and medical data of the sample are presented in Table 3.4.

The families were predominantly Caucasian and English speaking. Forty five per cent of families came from the higher social classes. Thirty per cent of the CF patients were under the age of 5 years, approximately 50% were aged between 6 and 12 leaving slightly less than 20% in the older age bracket of 13 to 22 years. Almost 80% of the CF patients obtained Shwachman Scores within the excellent to mild range. Only 12 patients (approximately 22%) were considered to be moderately to severely impaired by their illness. The generally good health of the CF patients was further supported by the finding that almost two-thirds of the CF patients had not been hospitalized during the study period. CF patients attended clinic on an average of once

TABLE 3.4

SOCIODEMOGRAPHIC AND MEDICAL CHARACTERISTICS OF THE STUDY SAMPLE[1]

CHARACTERISTICS		n	%
Social Class:[2]	I	10	16.67
	II	17	28.33
	III	21	33.00
	IV	8	13.33
	V	4	6.67
Race:	Caucasian	40	66.67
	Mixed	20	33.33
Language:	English	35	58.33
	Afrikaans	25	41.67
Shwachman Score:[3]	100-86 (excellent)	15	27.27
	85-71 (good)	14	25.45
	70-56 (mild)	14	25.45
	55-41 (moderate)	9	16.36
	40 and below (severe)	3	5.45

	MEAN	SD	RANGE
Age of patients (years)	8.46	4.87	1-22
Period since Diagnosis (months)	77.90	55.99	13-239
Number of Days in Hospital[4]	11.23	21.76	0-129
Number of Clinic Attendances[4]	15.42	7.26	3-38

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[1] n = 60 families

[2] Based on British Registrar General's Classification of Occupations

[3] Data missing for 5 patients

[4] Between January 1, 1984 and June 30, 1986

every two months. At the time of completing the study questionnaires, CF had been diagnosed for more than 5 years in over 50% of the families. Only 8 CF patients (13.33%) had been diagnosed for less than two years.

The sociodemographic and medical characteristics of CF patients and their siblings who completed study questionnaires are presented in Table 3.5. (The inclusionary criteria for the selection of patients and siblings were presented in Chapter 2). Eighteen CF patients (30% of the total number of CF patients) and 29 siblings (43.28% of the total number of siblings) completed questionnaires. Males and females were evenly distributed in the CF patient group, whereas there was a slight predominance of girls (55.17%) in the sibling group. Both groups were also predominantly Caucasian, English-speaking and had middle- or upper-class backgrounds. The mean ages of CF patients and siblings were approximately 14 and 15 years respectively. This also meant that on an average, siblings had received an extra year's education.

For only 1 CF patient and 2 siblings was the period since diagnosis less than 2 years. The average period since diagnosis was 11 years for patients and approximately 7 years for siblings. Two CF patients (11.11%) were only children.

A comparison was also made according to sociodemographic and medical variables, between families who did not have a child taking part in the study (n = 26) and families whose older children were included (n = 34). As was to be expected the latter sample included significantly more older children (mean

TABLE 3.5

SOCIODEMOGRAPHIC AND MEDICAL DATA OF CF PATIENTS AND SIBLINGS WHO COMPLETED QUESTIONNAIRES[1]

		PATIENTS		SIBLINGS	
		n	%	n	%
Gender:	Male	9	50.00	13	44.83
	Female	9	50.00	19	55.17
Language:	English	12	66.67	22	75.86
	Afrikaans	6	33.33	7	24.14
Race:	Caucasian	15	83.33	18	62.07
	Mixed	3	16.67	11	37.93
Social Class[2]:	I	4	22.22	5	17.24
	II	6	33.33	6	20.69
	III	4	22.22	14	48.28
	IV	3	16.67	4	13.79
	V	1	5.56	-	-
Shwachman Score:					
100-86 (excellent)		3	16.67		
85-71 (good)		3	16.67		
70-56 (mild)		4	22.22		
55-41 (moderate)		6	33.33		
40 or below		2	11.11		

	PATIENTS			SIBLINGS		
	MEAN	SD	RANGE	MEAN	SD	RANGE
Age (years)	14.22	4.1	9-22	15.45	3.33	10-23
Education (years)	7.67	3.1	4-12	8.60	2.98	3-15
Period since Diagnosis (months)	138.06	57.58	23-239	104.17	63.24	13-239
Number of days in Hospital[3]	17.11	16.77	0-41			
Number of Clinic Attendances[3]	17.06	8.77	3-38			

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1. n = 18 patients n = 29 siblings
2. Based on British Registrar General's Classification of Occupation
3. Between January 1, 1984 and June 30, 1986

14.38 versus mean 6.05 years, $p = 0.0001$), patients with significantly lower average Shwachman Scores (66.47 versus 75.71, $p = 0.0109$) and who had spent significantly longer average periods in hospital (16.53 versus 8.9 days, $p = 0.0009$). The Wilcoxon Sign Rank Test was used to compute these differences. The groups also differed significantly according to race. A significantly larger proportion of Caucasian children took part in the study (48.3% versus 8.32%, $Chi^2 = 6.33$, $p = 0.012$). The groups did not differ significantly on language, social class or number of clinic attendances.

3.4.2 Results of the CF Knowledge Test

Medical knowledge and common misconceptions about CF were assessed by 63 multiple choice questions. Completed tests were received from 60 mothers, 54 fathers, 18 patients and 29 siblings.

Knowledge scores were obtained by summing the number of correct responses on each item. COMPOSITE knowledge scores, reflecting overall test performance, and 9 SUB-SECTION scores reflecting the dimensions of this knowledge, were calculated for each family grouping. Sub-section analyses were undertaken to accurately identify specific areas of knowledge among family members since respondents could well have obtained the same composite scores yet have different gaps in this knowledge. A REVISED knowledge score which excluded the sub-sections of Terminology and General Medical Knowledge, was also calculated. This score thereby excluded the sub-sections containing items 17 and 31 which the panel of CF doctors had initially felt might not be correctly answered by the parents (Section 3.3.1.1). Parents were therefore expected to be able to answer all the questions appearing on the REVISED test.

Because the samples of fathers, mothers, patients and siblings were not independent, McNemar Chi square and Wilcoxon Sign Rank Tests were used for between-and-within-group comparisons of categorical and continuous data respectively (Siegel 1956). The 6 single mothers were excluded from statistical comparisons and correlations involving matched husband-wife pairs. Sample numbers used for the matched pair analyses are given in Table 3.6.

TABLE 3.6

SAMPLE NUMBERS FOR MATCHED PAIR ANALYSIS OF SCORES ON THE CF KNOWLEDGE TEST

PAIRED GROUPINGS	n
Mothers and Fathers	54
Patients and Siblings[1]	13
Patients and Mothers	18
Patients and Fathers	16
Siblings and Mothers	21
Siblings and Fathers	20

1 Where data were available from more than one sibling per family, an average score was used.

The findings on the CF Knowledge Test are presented as follows:

Composite Test Score Performance

Mean Sub-Section Performance

Individual Sub-Section Performance

3.4.2.1 Composite Scores on the CF Knowledge Test

Fathers and Mothers

The overall test performance of fathers and mothers is presented in Table 3.7. (A detailed summary of findings on the CF Knowledge Test for married mothers as a separate group is presented in Appendix 9). Average composite scores of 73.11% and 73.65% were obtained for fathers and mothers respectively. Paired analysis revealed a slightly, yet significantly, higher performance on the part of married mothers (76.29% vs 73.11%, p = 0.0245). Seven mothers (12.96%) and 5 fathers (9.26%) received scores of 90% or more, with over 50% of mothers receiving scores above eighty per cent. The median percentage for fathers was 73.81 per cent. Towards the lower end of the spectrum, 19 (31.67%) mothers and 22 (40.7%) fathers obtained scores of less than seventy per cent. Scores of below 50% were found in less than 10% of parents.

Compared to their scores on the Composite Knowledge Test, both mothers and fathers performed significantly better on the Revised Test (p = 0.0001) and mean percentages of 76.04 and 75.93 were were obtained respectively. The difference between the scores of matched pairs of mothers and fathers on the Revised Test was not statistically significant.

TABLE 3.7

MEANS, SD'S AND RANGES (RAW SCORES) ON THE CF KNOWLEDGE TEST ACCORDING TO FAMILY GROUPING

CF KNOWLEDGE TEST	n	MEAN	SD	RANGE
Composite Test (63 items)				
Fathers	54	46.06	8.58	26-62
Mothers	60	46.40	10.71	11-62
Patients	18	45.67	8.64	30-60
Siblings	29	40.44	10.32	11-56
Revised Test (56 items)				
Fathers	54	42.52	7.12	26-55
Mothers	60	42.58	9.21	11-55
Patients	18	42.33	7.08	30-53
Siblings	29	37.72	9.19	11-51

In view of an expected performance of one hundred per cent on the Revised Test, the panel of CF clinic doctors had overestimated the CF medical knowledge of mothers and fathers by approximately 24 per cent.

The test performances of mothers and fathers were moderately correlated on both the Composite Knowledge Test ($r_s = 0.63$, $p = 0.0001$) and the Revised Knowledge Test ($r_s = 0.60$, $p = 0.0001$).

Patients and Siblings

Patients and siblings obtained average Composite Test Scores of 72.49% and 64.19% respectively. Average Revised Test Scores for patients and siblings were alternatively 75.59 and 67.36 per cent. Paired analysis revealed that patients were significantly better informed than siblings on both the Composite and Revised Knowledge Tests ($p = 0.0309$ and $p = 0.0221$ respectively).

The overall test performances of patients and siblings were also examined in relation to those of their parents. The Test score differences between patients and mothers and patients and fathers failed to reach statistical significance on either the Composite or the Revised Knowledge Test. In contrast mothers were significantly more knowledgeable than siblings on both the Composite ($p = 0.004$) and the Revised Knowledge Test ($p = 0.0001$). Fathers, too, were significantly better informed than siblings on the Composite ($p = 0.001$) and the Revised Knowledge Test ($p = 0.0021$).

3.4.2.2 Mean Sub-Section Scores on the CF Knowledge Test

Frequency data (means, SD's and ranges) on the 9 sub-sections are presented in Appendix 10 for each family grouping. Henceforth, however, tests scores will be presented in the form of percentages.

Fathers and Mothers

Parental performance on each of the sub-sections is presented in Table 3.8.

TABLE 3.8

MEAN SUB-SECTION SCORES (% CORRECT RESPONSES) ON THE CF KNOWLEDGE TEST ACCORDING TO FAMILY GROUPING[1]

SUB-SECTION	NO OF ITEMS[2]	FATHERS	MOTHERS	PATIENTS	SIBLINGS
General CF Knowledge	13	83.90	83.97	80.34	73.74
Respiratory Symptomatology and Treatment	5	69.63	74.33	67.78	60.69
Physiotherapy	7	87.04	90.43	83.33	72.44
GIT Symptomatology and Treatment	5	83.40	81.67	90.00	73.10
Nutrition	13	71.79	71.41	69.66	62.07
Genetics	6	69.75	66.67	47.22	44.83
Anatomy	7	84.13	85.48	89.70	80.30
Terminology	3	43.31	46.11	42.59	29.88
General Medical Knowledge	4	56.02	60.83	51.39	45.69

[1] n = 54 fathers n = 60 mothers n = 18 patients n = 29 siblings

[2] Number of items in each sub-section

Parents received combined scores of over 80% on the four sub-sections of physiotherapy, general CF knowledge, anatomy and GIT symptoms and treatment. They were, however, less informed about respiratory symptoms and treatment, nutrition and genetics. Indeed, the CF doctors had overestimated parental performance on these sub-sections by almost thirty per cent. Parental performance was lowest on the sub-sections of terminology and general medical knowledge. With the exception of items 17 and 31, the panel of CF doctors had once again misjudged parental knowledge in these areas. Differences between the scores of fathers and mothers were not significant for any of the sub-sections.

Patients and Siblings

In keeping with parental trends, patients and siblings performed better on the sub-sections relating to physiotherapy, general CF knowledge, GIT symptomatology and treatment and anatomy (Table 3.8). Patients, too, obtained mean scores of over 80% on these sub-sections. Patient and sibling knowledge on genetics, general medical information and terminology was, however, particularly weak. The performance of patients was significantly superior to that of siblings on the sub-sections of physiotherapy ($p = 0.0209$), GIT symptoms and treatment ($p = 0.0215$) and nutrition ($p = 0.0016$).

The sub-section knowledge of patients and siblings was also compared with that of their fathers and mothers. Sub-section differences between mother-patient and father-patient pairs failed to reach statistical significance. Mothers were, however, significantly better informed than siblings on general CF knowledge ($p = 0.0055$), respiratory symptomatology and treatment ($p = 0.0102$) and genetics ($p = 0.0282$). Fathers obtained proportionately higher scores than siblings on general CF knowledge ($p = 0.0458$), terminology ($p = 0.0396$) and general medical knowledge ($p = 0.0135$). It was noteworthy that patients obtained higher mean scores than parents on the GIT and anatomy sub-sections.

3.4.2.3 Individual Sub-Section Performance on the CF Knowledge Test

In order to accurately identify areas of high and low knowledge, a comprehensive analysis of each sub-section was undertaken for fathers and mothers (Table 3.9) and patients and siblings (Table 3.10). The remainder of this section is based on findings presented in these Tables. (For an exact description of item content, the reader is referred to pp 3 - 11 of the original questionnaire in Appendix 1. Note: Because pp 3-11 were exactly the same for all family groupings they have been included in the parental questionnaire only).

a. General CF Knowledge

Fathers and Mothers

This was the most heterogeneous sub-section and included such assorted topics as aetiology, diagnosis, general symptomatology and principles of management. Not surprisingly well over 90% of parents knew that the lungs and pancreas are the main organs affected by CF, that thick and sticky mucus is an important determinant of frequent chest infections, that the sweat test is the most accurate way to diagnose CF, that smoking is detrimental to the health of the CF child and that the illness is lifelong. Although the percentage bordered on 90% for items 55 and 56, it is nevertheless disturbing that even very small percentages of parents erroneously believed that a child could outgrow CF and that marriage was contraindicated for CF persons. Almost 13% of parents incorrectly believed

TABLE 3.9

CF KNOWLEDGE (% CORRECT RESPONSES) FOR FATHERS AND MOTHERS ACCORDING TO SUB-SECTION AND ITEMS[1]

GENERAL CF KNOWLEDGE			RESPIRATORY SYMPTOMS AND TREATMENT			PHYSIOTHERAPY			GIT SYMPTOMS AND TREATMENT			NUTRITION		
ITEM NO[2]	FATHERS	MOTHERS	ITEM NO	FATHERS	MOTHERS	ITEM NO	FATHERS	MOTHERS	ITEM NO	FATHERS	MOTHERS	ITEM NO	FATHERS	MOTHERS
1	98.15	96.67	6	92.59	96.67	19	88.89	91.67	28	92.59	85.00	34	48.15	53.33
2	94.44	83.33	12	74.07	83.33	20	85.19	85.00	29	92.59	85.00	35	79.63	68.33
3	94.44	96.67	13	61.11	75.00	21	85.19	88.33	30	77.78	83.33	36	55.55	51.64
4	87.04	86.67	15	87.04	88.33	22	98.15	93.33	32	75.93	80.00	37	75.93	66.67
5	94.44	95.00	26	33.33	30.00	23	96.30	96.67	33	77.78	75.00	38	94.44	91.67
7	81.48	83.33				24	85.19	93.33				39	48.15	53.33
10	53.70	56.67				25	70.37	85.00				40	87.04	91.67
14	25.93	21.67										41	83.33	78.33
27	92.59	91.67										42	83.33	85.00
47	85.19	90.00										43	70.37	71.67
48	98.15	93.33										44	74.07	78.33
55	90.74	96.67										45	40.74	45.00
56	94.44	90.00										46	92.59	93.33

GENETICS			ANATOMY			TERMINOLOGY			GENERAL MEDICAL		
ITEM NO	FATHERS	MOTHERS	ITEM NO	FATHERS	MOTHERS	ITEM NO	FATHERS	MOTHERS	ITEM NO	FATHERS	MOTHERS
49	79.63	86.67	57	98.15	96.67	8	31.48	35.00	11	53.70	55.00
50	70.37	70.00	58	88.89	88.33	9	42.59	46.67	16	72.22	85.00
51	66.67	65.00	59	72.22	80.00	31	55.55	56.67	17	42.59	43.33
52	77.78	76.67	60	83.33	81.67				18	55.56	60.00
53	53.70	40.00	61	74.07	80.00						
54	70.37	61.67	62	85.19	86.67						
			63	87.04	85.00						

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[1] n = 54 fathers n = 60 mothers

[2] Item numbers as these appear on original questionnaires

TABLE 3.10

CF KNOWLEDGE (% CORRECT RESPONSES) FOR PATIENTS AND SIBLINGS ACCORDING TO SUB-SECTION AND ITEMS[1]

GENERAL CF KNOWLEDGE			RESPIRATORY SYMPTOMS AND TREATMENT			PHYSIOTHERAPY			GIT SYMPTOMS AND TREATMENT			NUTRITION		
ITEM NO[2]	PATIENTS	SIBLINGS	ITEM NO	PATIENTS	SIBLINGS	ITEM NO	PATIENTS	SIBLINGS	ITEM NO	PATIENTS	SIBLINGS	ITEM NO	PATIENTS	SIBLINGS
1	98.15	96.55	6	77.77	82.76	19	94.44	65.52	28	94.44	68.97	34	88.89	44.83
2	88.89	82.76	12	94.44	72.41	20	77.22	62.07	29	94.44	86.20	35	72.22	44.83
3	66.67	72.41	13	61.11	37.93	21	77.78	86.21	30	88.89	65.52	36	38.89	51.72
4	55.56	62.07	15	88.89	86.21	22	94.44	93.10	32	88.89	72.41	37	72.22	55.17
5	100.00	86.21	26	16.67	24.14	23	94.44	79.31	33	83.33	72.41	38	100.00	82.76
7	83.33	65.52				24	66.66	65.52				39	33.33	41.38
10	55.56	44.43				25	83.33	55.17				40	77.78	89.65
14	50.00	24.14										41	83.33	82.76
27	88.89	89.65										42	88.88	86.20
47	94.44	75.86										43	83.33	65.52
48	83.33	86.21										44	77.78	55.17
55	94.44	82.76										45	5.56	20.69
56	88.89	89.65										46	83.33	86.20

GENETICS			ANATOMY			TERMINOLOGY			GENERAL MEDICAL		
ITEM NO	PATIENTS	SIBLINGS	ITEM NO	PATIENTS	SIBLINGS	ITEM NO	PATIENTS	SIBLINGS	ITEM NO	PATIENTS	SIBLINGS
49	55.56	68.97	57	100.00	93.10	8	61.11	37.93	11	38.89	41.38
50	66.66	31.03	58	94.44	82.76	9	27.78	13.79	16	77.77	72.41
51	50.00	48.28	59	83.33	79.31	31	38.88	37.93	17	33.33	31.03
52	38.89	51.72	60	83.33	68.97				18	55.56	37.93
53	38.89	31.03	61	77.78	68.97						
54	33.33	37.93	62	94.44	82.76						
			63	94.44	86.21						

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[1] n = 18 patients   n = 29 siblings

[2] Item numbers as these appear on original questionnaires

that CF was caused by bad lungs. Almost 20% of fathers and 10% of mothers were unaware that excessive mucus in the lungs was likely to result in a decreased appetite in the CF child.

The lowest scores received in this sub-section related to two commonly held misconceptions (items 10 and 14). Only about 55% of parents were aware that asthma and allergies often occurred in CF children. The common and frequent occurrence of chest infections in CF may well have influenced parental responses to item 14 since as many as 70% of parents erroneously believed that CF children are more susceptible to the common cold than their siblings. When scores on these two items were removed from the mean total percentages on this sub-section, total averages increased by almost 10 per cent.

There were no significant differences between husband and wife pairs on any of the items in this sub-section.

Patients and Siblings

Patient and sibling knowledge in this sub-section was erratic. Most respondents were aware of the lifelong nature of the illness (item 55), the important roles of mucus (items 2, 5, 6 and 7) and smoking (item 56) in increasing disease morbidity and they knew the major organs affected by the illness (item 1). Both groups were, however, far less certain about either the manner of diagnosis (item 3) or causation (item 4) of the illness.

Approximately 40% of patients and siblings incorrectly believed that CF is caused by bad lungs while a further 30% erroneously thought that CF was diagnosed by means of a chest X ray. The high visibility of chest pathology in CF may have influenced these answers. Matched pair analyses showed that mothers were significantly better informed than patients ($Chi^2 = 5.00$, $p = 0.0253$) and siblings ($Chi^2 = 4.00$, $p = 0.0455$) on item 3 (sweat test). Fathers, in turn, were significantly better informed than patients as to the cause of cystic fibrosis ($Chi^2 = 6.00$, $p = 0.0143$).

As happened with parents, patients and siblings were unclear as to the frequency of occurrence in the CF patient of asthma, allergies or the common cold (items 10 and 14). Surprisingly, patients were slightly, yet not significantly, better informed than their parents as to their susceptibility to the common cold. Guessing may have contributed to this finding. Mean total percentages of patients and siblings increased by 5 and 7 per cent respectively when items 10 and 14 were removed from overall calculations on this sub-section.

The majority of CF patients knew that they might need extra salt during hot spells (item 47). Its importance was also recognized by almost three-quarters of the siblings. Mothers were significantly better informed on this item than siblings ($Chi^2 = 6.00$, $p = 0.0143$).

b. Respiratory Symptomatology and Treatment

Fathers and Mothers

Increased amounts of mucus and discoloured sputum are classical symptoms of bacterial chest infections in CF (Goodchild and Dodge 1985). It was, therefore, not surprising that most parents correctly answered item 6. Almost 13% of mothers and 26% of fathers, however, failed to associate green sputum with bacterial chest infections (item 12). The difference between parental scores on this item was statistically significant ($Chi^2 = 4.46$, $p = 0.0348$).

Although cough mixtures are commonly prescribed to dry up excess mucus in 'healthy' children, their use is generally contraindicated in CF (Goodchild and Dodge 1985). Significantly more fathers than mothers (38.89% versus 24.07%, $Chi^2 = 4.765$, $p = 0.0290$) incorrectly believed that cough mixtures had a useful role to play in the treatment of CF (item 13). Only one-third of parents was aware that nebulized antibiotics should be administered after physiotherapy (item 26). One-third of parents incorrectly answered this question, while the remaining one-third indicated that they did not know the answer.

Patients and Siblings

Like their parents, patients and siblings were similarly well-informed regarding the significance of mucus in chest infections (items 6 and 12). The slightly poorer performance of patients on item 6 (mucus increases during a chest infection) may well be ascribed to the fact that not all CF children produce large quantities of mucus during chest infections. In contrast, the need for regular sputum testing during clinic visits may well underscore the importance of colour changes and thus contributed to higher scores on item 12. The majority of patients and siblings recognized that CF is not infectious (item 15). Although patients and siblings were well-informed as to the symptomatic connotations of mucus in chest infections, they were far less knowledgeable about how to treat chest infections. Almost 40% of patients and 60% of siblings misconstrued the usefulness of cough mixtures in treatment (item 13). Even more alarming was the finding that 72.22% of patients mistakenly believed that antibiotics should be administered before physiotherapy (item 26). Siblings were, not surprisingly, equally misinformed. Parents, too, had displayed ignorance on this important treatment issue.

Matched pair analysis revealed no statistically significant differences between parents and patients and parents and siblings in this sub-section.

c. Physiotherapy

Fathers and Mothers

The removal of secretions by regular chest physiotherapy is an integral part of most CF treatment regimens (Stern 1986). Regular exercise is also encouraged by most CF clinics. Most parents were well-informed about these aspects of treatment and the highest proportion of correct responses for mothers and fathers alike was recorded in this sub-section. Mothers obtained scores of over 90% on 5 out of 7 items, with fathers averaging well over 80 per cent.

The important role of chest physiotherapy and exercise as a means for loosening and removing harmful secretions (items 20, 22, 23) was well knew by the majority of parents. Most parents recognized that physiotherapy should continue everyday even if the CF child were well (item 21) and that it should be administered before breakfast in both the young and older patient (items 24 and 25). In general mothers were better informed as to the most effective timing of physiotherapy, with a significantly larger proportion correctly answering item 25 (87.03% versus 70.37%, $Chi^2 = 4.26$, $p = 0.0334$). This latter finding was not entirely unexpected as fathers are likely to become less involved in the daily routine of physiotherapy with increasing patient age.

Patients and Siblings

Almost three-quarters of the patients knew that the purpose of physiotherapy was to remove harmful secretions (item 20). Whereas over 80% of patients realized that older CF children should perform their physiotherapy before breakfast, not unexpectedly, they were far less certain as to the timing of physiotherapy for smaller children (items 25 and 24 respectively). Patients were significantly better informed than their siblings as to the timing of physiotherapy for older patients (Chi^2 = 4.00, p = 0.0455).

It was interesting to note that although almost 95% of patients recognized that physiotherapy should continue everyday even when a chest infection was being treated with antibiotics (item 19), over 20% did not believe in its value when a child was well (item 21). It was also not entirely unexpected that siblings would be less informed than patients about the need for continuous physiotherapy (item 10) (Chi^2 = 5.00, p = 0.0253). The majority of patients and almost 80% of siblings recognized that exercise was a useful means of loosening mucous secretions (item 23).

None of the differences in scores between matched patient-mother or patient-father pairs were statistically significant. In contrast, significantly more siblings than their fathers were not aware of the need for continuous physiotherapy (item 19) (Chi^2 = 4.00, p = 0.0455) and significantly more mothers than siblings (Chi^2 = 7.00, p = 0.0082) knew that small children should have their physiotherapy before breakfast (item 24). In addition, significantly fewer mothers than siblings (Chi^2 = 4.5, p = 0.0339) incorrectly believed that physiotherapy could cure lung infections (item 20).

d. Gastrointestinal Symptomatology and Treatment

Fathers and Mothers

Pancreatic enzyme replacement is necessary in at least 90% of CF children (Goodchild and Dodge 1985). Four out of five items in this sub-section related to the function and administration of pancreatic enzymes. A high proportion of both fathers and mothers knew that CF children have too few pancreatic enzymes (item 29) and that an important function of which is the digestion of food (item 28). Less than 10% of parents were unable to answer these items. Although a fairly high percentage of mothers (87.04%) and fathers (77.78%) realized that oily stools were not a side-effect of pancreatic supplementation (item 30), it was nevertheless disturbing that 13% of mothers and 22% of fathers either incorrectly answered or were unable to answer this item.

Because of their implications for effective treatment, items 32 (enzymes must be taken with ALL meals) and 33 (best time to take pancreatic enzymes is before or during meals) can be considered critical knowledge. Yet between 15% and 25% of mothers and 25% of fathers failed to answer these items correctly.

Patients and Siblings

Approximately 95% of patients knew the reasons for, and functions of, pancreatic supplementation (items 28 and 29). In fact, patients scored consistently although not significantly, higher than their mothers on all 5 items in this sub-section. Only one patient mistakenly believed that oily stools result from the side-effects of pancreatic supplementation, while another was uncertain (item 30). Whereas well over 80% of siblings knew that CF children have too few pancreatic enzymes (item 29) they were, not surprisingly, less informed about the frequency and timing of this form of treatment. This is understandable considering the highly individualized nature of pancreatic supplementation.

Matched pair analysis indicated that significantly more patients than siblings ($Chi^2 = 5.00$, $p = 0.0253$) related oily stools to levels of fat in the diet. There were no significant differences in this sub-section between the scores of CF patients or siblings and those of their parents.

e. Nutrition

Fathers and Mothers

Adequate nutrition is an important adjunct to the overall management of CF (Soutter, Kristidis, Gruca and Gaskin 1986). Knowledge of nutrition was, however, variable particularly with respect to its application to specific treatment problems such as the need to increase energy or weight. Only 13% of mothers and 16.7% of fathers were correct on all items in this sub-section.

Knowledge of general nutritional facts was reasonable. A high proportion of parents knew that skim milk and jam are low in fat (items 38 and 40), that butter and cheese are high in fat (item 42) and that spaghetti, rice and porridge are examples of carbohydrates (item 41). It was, nevertheless, not anticipated that more fathers than mothers (79.63% versus 72.22%) would know that calories measured the energy, as opposed to vitamin, content of foods. Fathers again scored better than mothers on item 37 which identified *caloreen as a useful source of carbohydrate (75.93% versus 70.37%).

While carbohydrates and proteins can both be considered sources of calories, these are far more readily available to the CF child in the form of carbohydrates. This distinction was not obvious to many parents, almost 40% of whom believed protein to be a better source of calories (item 36). Less than 15% of parents acknowledged that they did not know the correct answer.

The application of nutritional knowledge produced mixed findings. Whereas more than 70% of parents recognized that a slice of fruit cake, as compared to one large apple, would lead to a greater weight gain (item 44), somewhat fewer mothers (57.41%) and fathers (48.15%) recognized that a medium-sized banana would contain more energy than 2 carrots (item 39).

Whereas until recently low fat diets were considered to be an important adjunct to controlling the gastrointestinal symptoms of CF, they are no longer advocated because of the improved potency of new pancreatic enzyme replacements (Dodge 1985). Thus for the majority of CF children, dietary manipulation should be kept to a minimum (Goodchild and Dodge 1985). Items 34 and 45 tested family members' knowledge on this issue. Almost 50% of parents mistakenly believed that fats should be excluded from the CF child's diet. Less than 10% of parents indicated that they did not know the answers, reflecting their firm, yet mistaken, conviction that fats should be excluded. Less than 8% of parents expected a low fat diet to cure the symptoms of CF (item 46). Considering their vital role in the management of steatorrhoea, it was alarming that approximately 20% of mothers and 30% of fathers were unaware of the relationship between the dosage of pancreatic enzymes and the amount of fat in the diet (item 43).

No significant differences between husband and wife pairs were found in this sub-section.

Patients and Siblings

Like their parents, patients and siblings were well-informed regarding the main constituents of familiar foodstuffs (items 38, 40, 41 and 42). With the exception of item 40, where almost 23% of patients wrongly thought jam contained more fat than peanut-butter, performances reflected scores well over 80% on these items. They also showed similar areas of ignorance to parents, when applying nutritional principles to actual eating patterns. Less than 40% of patients selected carbohydrates over fats as a better source of calories for the CF child (item 36). Even siblings were better informed on this item (38.89% vs 51.72%), although guessing may have influenced this finding. Significantly more patients than siblings recognized that calories measure the amount of energy in food (72.22% vs 44.83%, Chi^2 = 4.00, p = 0.0455). Given this latter result it was surprising that only one-third of the patients correctly identified a medium-sized banana as a better source of energy than 2 raw carrots (item 39). Siblings were only slightly better informed than patients on this item (41.38% vs 33.33%). Respondents may well have been misled by the 'quantities' i.e. Two carrots as opposed to ONE banana, instead of taking cognizance of the calorific content. Patients did, however, perform better on item 44 which required that respondents identify which foodstuffs (an apple or a slice of fruit cake), if eaten regularly, would lead to a greater weight gain. Almost 80% of patients correctly answered this question, compared to only 55.17% of siblings. This difference was not, however, statistically significant.

It was to be anticipated that significantly more patients than siblings would recognize caloreen as being a useful source of carbohydrate (item 37) (Chi^2 = 5.00, p = 0.0253). Only 1 patient incorrectly stated that caloreen was a source of fat, while 5 (27.78%) did not know the answer. Patients were also slightly better informed about this than their mothers on this item (72.22% vs 66.67%). Fathers, in turn, were significantly better informed than siblings

($Chi^2 = 4.5$, $p = 0.0339$). Over 80% of patients were aware of the relationship between the dosage of pancreatic enzymes and the amount of fat in the diet (item 43). Two patients mistakenly linked dosage with the carbohydrate content of food and 1 patient did not know the answer. Approximately two-thirds of siblings were aware of this relationship.

The inconsistent pattern of patients' responses to items 34 and 45 reflected their considerable confusion regarding the role of fat in their diets. Whilst only 10% of patients advocated a completely fat free diet in response to item 34, over 80%, in turn, believed that fats should be completely excluded from the CF patient's diet (item 45). Although parents were similarly misinformed on this issue, their responses had nonetheless been consistent across these two items. This inconsistency is further underscored by the findings that patients scored significantly better than fathers ($Chi^2 = 6.4$, $p = 0.0114$) and mothers ($Chi^2 = 4.00$, $p = 0.0455$) on item 34, while mothers, in turn, scored significantly better than patients ($Chi^2 = 9.00$, $p = 0.0027$) on item 45. Siblings' responses to these items reflected similar levels of ignorance. Almost 70% of siblings believed that fats should be completely excluded from the CF patient's diet (item 45). Indeed, siblings were significantly more ignorant than both their fathers ($Chi^2 = 6.00$, $p = 0.0143$) and their mothers ($Chi^2 = 7.35$, $p = 0.0067$) in this regard.

f. Genetics

Fathers and Mothers

Items in this sub-section assessed knowledge of carrier status, risks of recurrence and prenatal diagnosis. Approximately 30% of parents correctly answered all 6 items, while almost one-third obtained scores of less than 30 per cent.

Knowledge of genetics was mixed with parents obtaining consistently poorer scores on items which required the application of genetic concepts. Whereas approximately 80% of parents knew that the CF gene had to be carried by both parents (item 49), and over three-quarters knew that this translated into a one in four chance of producing an affected child (item 52), considerably fewer parents could correctly estimate the chances of their NOT having a child with CF (item 53). Indeed, almost 40% of mothers and 31% of fathers gave themselves a mere 25% chance of not having an affected child. Judging from the poorer performance on item 53, parents seemed less able to calculate probabilities when these were expressed as percentages. Furthermore, a meaningful proportion of fathers and mothers either did not know (14.81% and 21.67% respectively) or incorrectly assessed (14.18% and 16.67% respectively) their chances of having a third affected child (item 54).

In spite of its relatively recent discovery, approximately 70% of parents correctly answered the question relating to amniocentesis in CF (item 50) and two-thirds of parents were aware of the reliability of the prenatal diagnosis (item 51). Score differences between mothers and fathers were not statistically significant on any of these items.

Patients and Siblings

Patients and siblings were poorly informed about basic and applied genetics and only 2 (11.11%) patients and 3 (10.34%) siblings managed to correctly answer all the items in this sub-section. Slightly more than half the patients knew that both parents had to carry the CF gene (item 49), while less than 40% were aware of the recurrence risks to future pregnancies (items 52, 53 and 54). Correspondingly low levels of genetic knowledge were recorded among siblings. More siblings than patients knew that both parents had to carry the CF gene (68.97% vs 55.56%) and that recurrence risks are 1 in 4 (51.72% vs 38.89%). These findings could have been the result of guessing.

It was not anticipated that as many as two-thirds of patients would know about amniocentesis (item 50). Patients' knowledge on this item was significantly better than that of their siblings ($Chi^2 = 6.00$, $p = 0.0143$). Approximately half the sample of patients and siblings were aware of the reliability of the prenatal diagnosis.

g. Anatomy

Fathers and Mothers

Family members had to identify 7 body parts on a simple line drawing. These included the lungs, heart, liver, stomach, pancreas, small and large intestines (items 57-63). Almost all fathers and mothers correctly identified the lungs, with a further 88.6% of parents correctly naming the heart. Mothers achieved scores of 80% or more on the remaining items, whereas fewer fathers correctly identified the pancreas (74.07%) and the liver (72.22%).

Patients and Siblings

Response patterns mirrored those of parents, with patients and siblings correctly identifying the heart, lungs and intestines more frequently than the other organs. Surprisingly, significantly more patients than siblings correctly identified the heart ($\text{Chi}^2 = 4.00$, $p = 0.0455$).

In spite of its pathophysiological significance in their disease, over 20% of patients (and 30% of siblings) failed to identify the pancreas. It is interesting to note that while the majority of all family members (approximately 97%) knew that the pancreas is a major organ involved in CF (item 1), they were clearly far less certain as to its location in the body.

h. Terminology

Fathers and Mothers

This sub-section included three medical terms which describe symptoms common to CF viz dyspnoea, haemoptysis and steatorrhoea. Knowledge of common CF terminology was uniformly low and this was further reinforced by the high percentage of 'I don't know' responses from parents. Less than 30% of parents managed to correctly identify all three terms. Parents seemed most familiar with the term 'steatorrhoea' where responses reached an average of over 50 per cent (item 31). Slightly more than 50% of mothers and 40% of fathers correctly associated haemoptysis with coughing up blood (item 9). Parental ability to relate dyspnoea with shortness of breath was even poorer, with only 35% of mothers and 33.33% of fathers correctly answering this question (item 8).

Patients and Siblings

Patients and siblings were similarly ignorant of the medical terms for common CF symptoms. Six (33.33%) patients and 12 (41.38%) siblings were unable to identify any of the technical terms. Curiously patients correctly linked dyspnoea with shortness of breath more frequently than their parents (item 8). They also performed significantly better than siblings on this item ($Chi^2 = 4.00$, $p = 0.0455$). Less than 30% of patients and 15% of siblings knew the medical term for coughing up blood (item 9) and less than 40% of both groups were aware of the meaning of steatorrhoea (item 31). Significantly fewer siblings than their fathers ($Chi^2 = 5.44$, $p = 0.0196$) and their mothers ($Chi^2 = 8.00$, $p = 0.0047$) knew the meaning of haemoptysis. There were no significant differences between patients and their parents on these items. The average proportions of 'I don't know' responses for patients and siblings on all 3 items were 35.18% and 45.98% respectively.

i. General Medical Knowledge

Fathers and Mothers

This sub-section included 4 general medical questions which have particular relevance to the management of CF. With the exception of item 16, concerning the 'normal' temperature, levels of general medical knowledge were low. Less than 20% of parents managed to correctly answer all the items in this sub-section.

A significantly larger proportion of mothers than fathers correctly identified a 'normal' temperature (87.04% vs 72.22%, $Chi^2 = 4.57$, $p = 0.0325$). Less than half the parents, however, knew that a child's temperature is usually highest in the late afternoon.

Knowledge of bacterial and viral infections was surprisingly low, with parents frequently confusing the two agents. Approximately 25% of fathers and mothers incorrectly believed that viral infections could be effectively treated with antibiotics while a further 18.42% of parents were uncertain. Mothers were slightly better informed in this regard. Similarly poor results were obtained on item 11 where over 40% of parents incorrectly linked the cause of the common cold to bacterial infection.

Although the panel of CF clinic doctors had anticipated a poor performance on item 17, they had expected that parents would be able to answer the remaining 3 items in this subsection. Clearly they were unaware of parental confusion about bacterial and viral infections.

Patients and Siblings

Patients and siblings performed even more poorly than their parents on items relating to bacterial and viral infections. Over half the sample of patients and siblings mistakenly thought that bacteria caused the common cold (item 11). Despite parents' own ignorance in this respect, significantly more mothers than patients ($Chi^2 = 4.00$, $p = 0.0455$) knew that common colds are caused by viruses, while fathers' scores were also significantly better than those of patients ($Chi^2 = 7.36$, $p = 0.0067$). Approximately 55% of patients and 40% of siblings knew that antibiotics were effective in the treatment of bacterial infections (item 18). Almost one quarter of both groups indicated that they did not know the answer. Differences between mother-sibling pairs on this item were statistically significant ($Chi^2 = 4.5$, $p = 0.0339$).

Whereas over 70% of patients and siblings correctly identified a 'normal' temperature (item 16), only one-third were aware that a child's temperature is highest in the late afternoon (item 17). Fathers and mothers scored significantly better than siblings on item 16 ($Chi^2 = 6.0$, $p = 0.0143$ and $Chi^2 = 4.5$, $p = 0.0339$ respectively).

3.4.3 Sociodemographic and Medical Correlates of CF Knowledge

The Composite CF Knowledge (i.e. total scores) of family members was examined in relationship to selected sociodemographic and medical variables (refer Section 3.3.2 for choice of variables).

Fathers and Mothers

The independent and interaction effects on Composite CF Knowledge of social class, race and language were assessed by means of bivariate and multivariate analysis of variance. Repeated measures analysis of variance was also used to examine the effect of parental status (i.e. father or mother) on CF Knowledge. Significant independent main effects on the CF Knowledge of fathers and mothers were found for social class ($F = 40.53$, df 4, $p = 0.0001$ and $F = 46.47$, df 4 and $p = 0.0001$ respectively), race ($F = 14.24$, $p = 0.0001$ and $F = 23.58$, $p = 0.0001$ respectively) and language. The effect of language was, however, significant for mothers only ($F = 6.56$, $p = 0.0131$). Subsequent pairwise testing (Gabriel's Test) revealed significantly superior CF Knowledge among fathers and mothers from social classes I and II and fathers and mothers of Caucasian origin ($p < 0.05$). Frequency data of fathers' and mothers' CF Knowledge according to social class, race and language are presented in Table 3.11. The mean performances of

TABLE 3.11

SUMMARY DATA (MEANS AND SD's) OF PARENTAL PERFORMANCE ON THE CF KNOWLEDGE TEST ACCORDING TO RACE, LANGUAGE AND SOCIAL CLASS

	CF KNOWLEDGE[1]								
	FATHERS			MOTHERS			PARENTS		
	n	MEAN	SD	n	MEAN	SD	n	MEAN	SD
RACE									
Caucasian	38	48.68	7.61	40	50.48	7.60	78	51.27	7.77
Mixed	16	39.81	7.65	20	38.25	11.53	36	40.50	10.38
LANGUAGE									
English	32	47.97	7.82	35	49.29	9.64	67	50.28	8.99
Afrikaans	22	43.27	9.06	25	42.36	11.01	47	44.43	10.42
SOCIAL CLASS									
I	10	55.60	4.93	10	56.50	4.99	20	56.05	4.85
II	16	50.56	6.29	17	54.65	3.74	33	52.67	5.46
III	20	44.45	7.76	21	46.14	7.78	41	45.32	7.72
IV	6	42.50	8.19	8	42.12	9.79	14	42.29	8.91
V	2	30.00	4.24	4	22.00	10.42	6	24.67	9.27

[1] Possible range of scores was 0 to 63

fathers and mothers showed a definite stepwise increase in CF knowledge according to social class.

No interaction or parental status effects were noted in any of these analyses. (F and p values are presented in Tables 3.12 and 3.13). Closer inspection of the former data revealed a marked social class effect throughout the various analyses. Although race, as an independent variable, had been a highly significant factor influencing fathers' and mothers' CF Knowledge, when entered together with social class, its effect became non-significant while that of social class remained highly significant. These findings suggested a far more powerful social class effect. Given that 90% (n = 18) of the families of mixed origin came from social classes III, IV and V, this finding was not unexpected. In similar vein, when entered alongside social class, the effect of language remained non-significant for fathers and became less significant for mothers ($p = 0.0245$). That language should have remained significant for mothers only may have resulted from the fact that more Afrikaans speaking mothers than fathers (6 and 4 respectively) fell into social classes IV and V and that no English speaking fathers fell into social class V. Race also proved to have a more powerful effect on CF Knowledge than language for fathers and mothers. Closer examination of the frequency data (Table 3.14) revealed that the lowest mean CF Knowledge scores were recorded among Afrikaans speaking families of mixed origin.

TABLE 3.12

STATISTICAL DATA (F AND p VALUES) ON THE INTERACTION EFFECTS OF RACE, LANGUAGE AND SOCIAL CLASS ON THE CF KNOWLEDGE OF FATHERS AND MOTHERS[1]

	FATHERS			MOTHERS		
	DF	F VALUE	p VALUE	DF	F VALUE	p VALUE
RACE and SOCIAL CLASS	7	5.90	0.0001*	7	9.75	0.0001*
Race	1	0.48	0.4923	1	1.83	0.1825
Social Class	4	4.57	0.0034*	4	5.80	0.0007*
Race x Social Class	2	1.20	0.3093	2	0.84	0.4392
LANGUAGE and SOCIAL CLASS	8	5.30	0.0001*	8	7.72	0.0001*
Language	1	2.87	0.0970	1	5.54	0.0245*
Social class	4	8.66	0.0001*	4	11.84	0.0001*
Language x Social Class	3	0.70	0.5554	3	0.43	0.7312
RACE and LANGUAGE	3	6.69	0.0007*	3	15.70	0.0001*
Race	1	15.31	0.0003*	1	36.93	0.0001*
Language	1	4.97	0.0302*	1	11.04	0.0017*
Race x Language	1	1.09	0.3017	1	3.89	0.0541
RACE, LANGUAGE and SOCIAL CLASS	4	10.94	0.0001*	4	19.66	0.0001*
Race	1	0.21	0.6494	1	2.78	0.1016
Language	1	0.50	0.4840	1	1.66	0.2043
Social Class	1	5.22	0.0267*	1	6.57	0.0135*
Race x Language x Social Class	1	0.38	0.5409	1	0.32	0.5768

1 n = 54 fathers n = 60 mothers

TABLE 3.13

F AND p VALUES (REPEATED MEASURES ANALYSIS OF VARIANCE) FOR THE EFFECTS OF PARENTAL STATUS ON CF KNOWLEDGE ACCORDING TO RACE, LANGUAGE AND SOCIAL CLASS

	DF	F VALUE	p VALUE
RACE			
Between Subject Effects			
Race	1	30.17	0.0001*
Within Subject Effects			
Parental Status	1	3.19	0.0798
Parental Status x Race	1	1.47	0.2303
LANGUAGE			
Between Subject Effects			
Language	1	5.56	0.0211*
Within Subject Effects			
Parental Status	1	3.65	0.0615
Parental Status x Language	1	0.23	0.6337
SOCIAL CLASS			
Between Subject Effects			
Social Class	4	66.86	0.0001*
Within Subject Effects			
Parental Status	4	1.00	0.3210
Parental Status x Social Class	4	0.09	0.7639

TABLE 3.14

SUMMARY DATA (MEANS AND SD's) ON THE CF KNOWLEDGE OF FATHERS AND MOTHERS ACCORDING TO RACE/LANGUAGE GROUPINGS

	COMPOSITE CF KNOWLEDGE					
	FATHERS			MOTHERS		
	n	MEAN	SD	n	MEAN	SD
Caucasian/English	23	51.26	7.43	23	54.39	7.84
Mixed/English	9	44.67	8.23	12	44.75	10.23
Caucasian/Afrikaans	15	48.53	8.07	17	49.47	6.98
Mixed/Afrikaans	7	37.14	6.26	8	32.38	10.99

When the effects of all three variables were considered together only that of social class remained significant for both fathers and mothers.

A clearer understanding of the magnitude of the effects of race, language and social class was obtained through an examination of their independent and combined contribution to the total variance in parental CF Knowledge scores. The R square statistic was calculated for this purpose. Since CF Knowledge showed a linear increase according to social class, this statistic was particularly useful for measuring explained variance (Kohout and Norwood 1981). It is, however, less sensitive to variance which is non-linear in nature. (The CF Knowledge Test scores of fathers and mothers were combined for these analyses in order to increase sample numbers and hence the power of this analysis). Findings for these analyses are presented in Table 3.15.

TABLE 3.15

PROPORTION OF VARIANCE IN PARENTAL CF KNOWLEDGE DETERMINED BY THE INDEPENDENT AND COMBINED EFFECTS OF RACE, LANGUAGE AND SOCIAL CLASS[1]

	DF	F VALUE	R^2	p VALUE
RACE	1	37.98	0.2532	0.0001*
LANGUAGE	1	10.27	0.0840	0.0018*
SOCIAL CLASS	4	31.31	0.5347	0.0001*
RACE and SOCIAL CLASS	5	26.22	0.5483	0.0001*
LANGUAGE and SOCIAL CLASS	5	28.24	0.5666	0.0001*
RACE, LANGUAGE and SOCIAL CLASS	6	25.08	0.5844	0.0001*

[1] n = 114 parents

The proportion of variance in parental CF Knowledge determined independently by race, language and social class was 25.32%, 8.4% and 53.47% respectively. When race and language were each combined with social class, this led to an increase in the proportion of total variance of only 1.36% and 3.19% respectively. When both race and language were combined with social class, the increase in total variance was 4.97 per cent. These findings confirmed that social class was an important predictor of parental performance on the CF Knowledge Test. Since the contribution of race and language to the total variance in parental CF Knowledge was negligible when combined with social class, it was decided to only include social class in remaining analyses in the study.

Although the 6 single mothers were shown to have significantly less CF Knowledge than their married counterparts, the small number of single parents made meaningful interpretation of these findings very difficult, the more so as social class was not associated with single parenthood. The author therefore elected not to analyze the remainder of the findings according to marital status. The role of marital status in understanding CF does, however, need further investigation but such research will need to take place among much larger numbers of single parents.

No significant relationships (Spearman rank correlation coefficients) were found between the CF Knowledge of parents and the age and medical characteristics (i.e. Shwachman score, period since diagnosis, days in hospital and number of clinic attendances) of the patients (p values: Appendix 11).

The effect of gender on parental CF Knowledge has already been documented (Tables 3.7 and 3.8). Mothers were only slightly more knowledgeable on the Composite Test while differences on the 9 sub-sections were not significant. As already noted the effect of parental status was non-significant when examined in relation to race, language and social class.

Patients and Siblings

Small sample sizes prevented the use of multivariate analysis to examine the combined effects of social class, race and language on the Composite Test performances of patients and siblings. Test score differences according to each of these variables were assessed independently by means of the Wilcoxon 2 Sample Test and Chi-square analysis. (For the purpose of these analyses social classes I and II and III, IV and V were combined). Test score differences according to social class were not significant for patients or siblings. The finding that siblings from social classes III, IV and V received proportionately higher scores than siblings from social classes I and II (67.02% vs 53.68%) was, however, contrary to parental findings. Test score differences according to race and language were also not significant for patients and siblings. Siblings of mixed origin and those who were Afrikaans speaking, however, obtained slightly higher scores (71.57% versus 63.78% and 71.24% versus 62.06% respectively). These findings were again counter to those reported for parents. Given their small cell numbers, these could well have been chance findings.

The Composite and Sub-Section Test Scores of patients and siblings were also examined according to the age and gender of the respondents (Table 3.16). With the exception of physiotherapy where scores were very close, older patients (14-22 years) scored consistently higher than younger patients. Older patients were significantly more informed on the sub-sections of general CF knowledge ($p = 0.0263$) and nutrition ($p = 0.0356$). Differences between the older and younger patients on

TABLE 3.16

MEAN SUB-SECTION SCORES ON THE CF KNOWLEDGE TEST (% CORRECT) FOR PATIENTS AND SIBLINGS ACCORDING TO GENDER AND AGE

	n	GENERAL CF INFORMATION	RESPIRATORY SYMPTOMS & TREATMENT	PHYSIOTHERAPY	GIT SYMPTOMS & TREATMENT	NUTRITION	GENETICS	ANATOMY	TERMINOLOGY	GENERAL MEDICAL
PATIENTS										
Male	9	76.08	62.22	80.95	86.67	59.85	38.83	88.89	33.33	38.89
Female	9	84.62	73.33	85.71	93.33	79.46	55.55	90.48	51.85	63.89
SIBLINGS										
Male	13	60.08	50.77	62.64	64.62	51.46	34.67	81.32	17.95	36.54
Female	16	70.38	67.50	82.14	80.00	70.46	53.17	80.36	37.50	54.69
PATIENTS										
9-13	9	73.54	64.44	84.13	88.89	61.54	38.83	85.71	37.04	44.44
14-22	9	87.15	71.11	82.54	91.11	77.77	55.55	93.65	48.15	58.33
SIBLINGS										
10-15	16	76.92	61.25	75.00	76.25	62.54	43.01	72.32	27.08	46.88
16-23	13	69.85	58.46	71.43	69.23	61.54	44.83	91.21	30.77	46.15

the Composite CF Knowledge Test (77.43% versus 67.55%) were, however, not significant. No differences, according to age, on either the Composite Test or Sub-Sections, were significant for siblings. It was, however, not anticipated that younger siblings (10-15 years) would obtain a mean Composite Test score superior to that of the older group (16-23 years) (66.48% versus 61.41%).

Female patients were consistently more knowledgeable than their male counterparts on all the sub-sections, although these differences were only significant in the case of nutrition ($p = 0.0442$). The superior performance of female patients on the Composite Test was not significant (77.84% versus 66.14%, $p = 0.0698$). In similar vein, female siblings out performed males on the Composite Test (70.63% versus 56.29%, $p = 0.0480$) and on 8 sub-sections. Sub-section differences were significant for nutrition ($p = 0.0284$) and GIT symptomatology and treatment ($p = 0.0344$).

The Composite CF Knowledge of male and female patients and siblings was also examined according to education and selected medical variables. With the exception of a strong positive correlation between Days in hospital and the CF Knowledge of male patients ($r_s = 0.81$, $p = 0.0008$), no other variables showed a significant relationship with CF Knowledge (p values: Appendix 12). Given the small number of male patients ($n = 9$), this finding must be treated with extreme caution.

3.4.4 Perception of Present and Future Health

Perception of Present and Future Health was assessed by means of two single items. As already noted, Perception of Future Health was used as an indirect indicator of 'knowledge' of prognosis in the present study. Response patterns, for all family groupings, on both sets of items were skewed in favour of positive assessments (Tables 3.17 and 3.18).

A substantial proportion of fathers (42.6%), mothers (45%) and siblings (51.72%) described the CF patient's Present Health as either excellent or very good. Whereas no parents assessed their CF child's Present Health as being poor or very poor, approximately 10% of siblings felt that the patient's Present Health was very poor. CF patients' own assessments of their Present Health were similar to those of parents, with almost 40% describing themselves as being in excellent or very good health. Only one patient felt that his health was poor.

Despite the progressive nature of the disease, only 20.75% of fathers, 22.8% of mothers and 10.34% of siblings predicted any deterioration in the CF patient's Future Health. In fact, only one mother indicated that in the future her affected child's health might be a great deal worse. Instead 47.17% of fathers, 38.5% of mothers and over 50% of siblings expected the affected child to be either cured or very much better in the future. Similarly, less than 20% of patients anticipated any worsening of their condition, while over 40% predicted an improvement.

TABLE 3.17

PERCEPTION OF THE CF PATIENT'S PRESENT HEALTH (% DISTRIBUTION) ACCORDING TO FAMILY GROUPING[1]

	FAMILY GROUPING			
	FATHERS	MOTHERS	PATIENTS	SIBLINGS
Excellent	5.56	11.67	5.56	17.24
Very good	37.04	33.33	33.33	34.48
Good	31.48	30.00	33.33	20.69
Fair	25.93	25.00	22.22	17.24
Poor	-	-	5.56	-
Very Poor	-	-	-	10.34

1 n = 54 fathers n = 60 mothers n = 18 patients n = 29 siblings

TABLE 3.18

PERCEPTION OF THE CF PATIENT'S FUTURE HEALTH (% DISTRIBUTION) ACCORDING TO FAMILY GROUPING[1]

	FAMILY GROUPING			
	FATHERS	MOTHERS	PATIENTS	SIBLINGS
Cured	16.98	17.54	11.11	20.69
Very much better	30.19	21.05	27.78	34.48
Little better	11.32	17.54	22.22	31.03
Remain the same	20.75	21.05	22.22	3.45
Little worse	16.98	17.54	5.56	10.34
Lot worse	3.77	3.51	11.11	-
Great deal worse	-	1.75	-	-

1 n = 53 fathers n = 57 mothers n = 18 patients n = 29 siblings

McNemar matched pair analyses revealed no significant differences between the family groupings concerning their perceptions of the CF patient's Present and Future Health. (For the purpose of these analyses perception of Present and Future health was dichotomized respectively as follows: excellent, very good, good/fair, poor, very poor and cured, very much better/little better, remain the same, little worse, lot worse, a great deal worse).

Perceptions of Present and Future Health were also examined in relation to selected sociodemographic and medical variables. Because there were no statistically significant differences between parental perceptions of Present and Future Health, their scores were combined for these analyses. None of these relationships were statistically significant (p values: Appendix 13). Chi-square analysis was used to examine the relationship between social class and Perception of Present and Future Health. Whereas social class was significantly associated with parental perception of Future Health ($Chi^2 = 20.83$, df 4, $p = 0.000$), its relationship with Present Health was not statistically significant. Parents from social classes III, IV and V were far more likely to expect their CF child's health to be cured or very much better in the future than was the case for parents from social classes I and II (30.7% versus 10.53%). Social class was not associated with CF patients' or siblings' perception of either Present or Future Health.

A more indirect measure of family members' outlook for the patient's future was obtained through their open-ended responses to the question: 'What worries you most about your child's illness/your illness/your brother or sister's illness?'

A content analysis of these individual responses according to family grouping revealed a far greater concern, and preoccupation with, the patient's future health than might have been suspected from the very positive attitudes gleaned from response patterns in Table 3.18. Indeed, fears about a limited lifespan, future deterioration and ultimate death were mentioned by approximately 40% of fathers. Only one father, however, actually used the word 'death' in his answer. Examples of verbatim comments from fathers included the following:
'It's incurable and the prognosis is not good'
'To have and love her, but not knowing for how long'
'His lifespan'
'The statistics'
'That he suffers a slow lingering deterioration that finally renders him bed-ridden and dies, in what should be the prime of his life'.

Approximately one-third of mothers expressed similar fears. Three mothers were able to use the word 'die' or 'death' in their responses, examples of which were:
'It's incurable'
'Having reared her to this stage, then having to lose her'
'That he'll suddenly deteriorate'.

With the exception of one patient who was worried about 'How and when I will die' and another who feared his 'lungs might conk in one day', no other CF patients expressed any fears about their future health. Instead their worries were characterized by here-and-now issues such as, inter alia, smelly stools, excessive thinness and stomach aches. In similar vein, when asked what worried their parents most, concerns about their future health were not mentioned by any patients.

In contrast, approximately 35% of siblings felt that their parents were worried about the patient's future health, the terminal nature of the disease and if a cure would ever be found. Slightly fewer siblings (31.03%) indicated that they personally were concerned about whether the patient's condition would deteriorate. Examples of their main worries included such comments as:

'It's a terminal sickness'
'I'm scared that he's going to die'
'I'm worried about his future, it won't be like mine'
'That he will never be cured'.

Despite these spontaneously expressed fears about the patient's future health, attributable mainly to the progressive and terminal nature of CF, only 16.67% of fathers, 10% of mothers, 6.67% of patients and 16% of siblings considered CF to be the worst illness a child could have (Table 3.19).

3.4 SUMMARY

In summary this chapter has reviewed the general literature which has assessed what chronically ill children and their parents understand about medical matters, together with the

TABLE 3.19

FAMILY MEMBERS' RESPONSES (% DISTRIBUTION) TO THE QUESTION: 'WHAT IS THE WORST ILLNESS A CHILD CAN HAVE?'

FAMILY GROUPING[1]							
FATHERS	%	MOTHERS	%	PATIENTS	%	SIBLINGS	%
Don't know	20.37	Cancer	31.67	Cancer	46.67	Cancer	48.00
Cancer	18.52	Don't know	21.67	Handicap	20.00	CF	16.00
Any terminal illness	18.52	Any terminal illness	16.67	Aids	13.33	Handicap	12.00
CF	16.67	Handicap	11.67	Not CF	13.33	Asthma	8.00
Handicap	14.81	CF	10.00	CF	6.67	Aids	4.00
Not CF	5.56	Lung infections	6.67	Other	12.00		
Other	5.56	Not CF	1.65				

[1] n = 54 fathers n = 60 mothers n = 15 patients n = 25 siblings

limited CF literature in this field. Recent research which has begun to evaluate knowledge as a potential explanatory variable was also touched upon. This was followed by the measurement and subsequent findings of a sample of family members' knowledge of CF, their perception of the patient's present and future health and the relationship between this knowledge and sociodemographic and medical variables.

Whereas this chapter has focused on current levels of disease-related knowledge among family members, the following chapter identifies areas about which they wish to increase their knowledge about CF. It also examines, inter alia, the degree to which they wish to be kept informed about CF and the main sources of their information about the illness.

CHAPTER 4

INFORMATION AND THE CF FAMILY

4.1 INTRODUCTION

This chapter is concerned with meeting the following aims of the study:

a. The assessment of information preferences, that is, how much family members want to be told about CF.

b. The assessment of information needs concerning selected medical and psychosocial topics and whether this information has been provided in the past.

c. The identification and evaluation of sources of CF information and guidance used by family members.

d. The identification of important sociodemographic and medical correlates of the above measures.

e. The examination of the relationship between CF knowledge and the quantity and quality of informational support.

f. The examination of the relationship between CF knowledge, information needs and personal and family adjustment.

4.2 REVIEW OF THE LITERATURE

The review begins by highlighting literature which has underscored the importance of information in facilitating a family's ability to cope with life-threatening illness. It is followed by writings and empirical research pertaining to the main sources of CF families' information about CF. Because the role of the doctor is particularly significant in information-giving, an historical account of changing attitudes towards disclosure within the medical profession has also been incorporated in the final section of the review.

4.2.1 Information-Seeking in Life-Threatening Childhood Illness

Following the diagnosis of a chronic childhood illness, families are faced with the task of learning as much as they can about the illness. Chesler and Yoak (1984) feel that the task of obtaining and assimilating large amounts of new information about an illness, its treatment and prognosis can create a significant source of 'intellectual stress' for family members. According to the coping model, information-seeking is a crucial coping skill in situations in which knowledge is limited (Feifel, Strack and Nagy 1987). The importance of this form of coping has, moreover, been underscored in several CF studies (Blumenthal 1969; McCollum and Gibson 1970 and McCubbin, McCubbin, Patterson et al 1983).

According to the literature, patients with a chronic life-threatening illness and their families engage in information-seeking for many different reasons. Futterman and Hoffman (1973) see this search for information as an attempt to gain intellectual mastery over the situation 'as though knowledge were actually power' (p.133). Yet other researchers feel that much information-seeking reflects parental efforts to check on the reliability of the doctors and the hospital (Friedman, Chodoff, Mason and Hamburg 1963 and Kirkpatrick, Hoffman and Futterman 1974). De Wet (1984) describes the reactions of several CF mothers in her sample who spent large amounts of time researching the illness to the extent that they saw themselves as 'more knowledgeable than the professionals' (p.169). In another example De Wet and Cywes (1985b) quote a CF parent as follows: 'I did not believe the doctors at all. I was sure they were talking nonsense. I proceeded to read and study cystic fibrosis - everything I could lay my hands on' (p.370).

Certain studies suggest that information-seeking is motivated by hope (Kirkpatrick, Hoffman and Futterman 1974 and Comaroff and Maguire 1981). De Wet and Cywes (1985b) cite the example of two couples who refused to accept a diagnosis of CF, a consequence of which they 'both undertook an avid search for more information on cystic fibrosis, if only to prove that a wrong diagnosis had been made in their case' (p.370).

Seeking new information may also serve to relieve parental feelings of guilt and responsibility for their child's illness (Chodoff, Friedman and Hamburg 1964) and to place their emotional reactions to the illness in perspective (Barbarin and Chesler 1986). In addition, parents are expected to be the frontline providers of information to siblings, relatives, friends and school personnel which, in turn, requires them to be well-informed about many aspects of the illness (Chesler and Barbarin 1984 and De Wet 1984).

Patients and parents also seek information to gain some measure of predictability over the future (Kirkpatrick, Hoffman and Futterman 1974). Several studies have identified an uncertain future and too little prognostic information as a significant source of stress for CF families (McCollum and Gibson 1970; Burton 1975; Falkman 1977 and De Wet 1984).

Sibinga and Friedman (1971) cite information for information-sake as an important reason for accumulating disease-related knowledge. They feel that 'being better informed can be a learning or growth-promoting experience for parents independent of any specific medical benefit to the child' (p.222).

Probably the single most important reason for seeking more information is so that parents and patients can obtain adequate and detailed information about the diagnosis, treatment and long-term management of the illness (Chesler and Yoak 1984).

While these studies have succeeded in drawing attention to the breadth of information-seeking in families faced with life-threatening illness, few have attempted to establish the content of the information being sought. To date, empirical research has not identified, with any degree of specificity, what family members want to know about CF. An important aim of the present study was to address this gap in the existing CF research.

4.2.2 Sources of Informational Support in CF

According to the coping model, information-seeking can be either facilitated or impeded by the quality and availability of informational support (Cohen and Lazarus 1980). In the face of chronic childhood illness the most frequently used sources of such support include the doctor (Burton 1975; Frydman 1981; Strauss and Wellisch 1981 and Phillips, Bohannon, Gayton and Friedman 1985), the family (Share 1972) and written information (Van Vechten, Satterwhite and Pless 1977). The remainder of the literature review focuses on the role of the family, written information and the medical profession as potential sources of disease-related information for CF families.

4.2.2.1 The CF Family as a Source of Informational Support

When a child has a serious and possibly fatal illness, parents are faced with the extremely difficult task of communicating these facts directly to the child, to his siblings and to each other. Accumulated evidence from a number of studies across various fatal illness groups (Burton 1975; Fife 1978; Buchannan, La Barbera, Roelofs and Olsen 1979; Obetz, Swenson, McCarthy et al 1980; De Wet 1984 and Fitzpatrick and Barry 1986) has identified the following as the most commonly reported parental difficulties: not knowing what to say to the affected child and his well siblings, particularly with respect to the prognosis; concern about the potential negative effects on the child of knowing the true nature of his condition and the parents' feelings of being ill-equipped to perform this task.

An early review article (Share 1972) has covered many of the controversies surrounding an open versus a closed approach to family communication in the face of fatal childhood illness. More recent empirical evidence, mainly from the fields of childhood cancer and CF, confirms earlier suggestions that honest and open communication within the family about the seriousness of a child's condition facilitates the child's adjustment (Spinetta and Maloney 1978; Slavin, O'Malley, Koocher and Foster 1982 and Smith, Gad and O'Grady 1983) and promotes more effective family functioning (Kaplan, Grobstein and Smith 1976; Falkman 1977; Spinetta, Swarner and Sheposh 1981 and

Davies, Spinetta, Martinson et al 1986). Evidence from the interview-based CF research also adds further support to the positive contribution which open and full communication makes to overall patient and family adjustment (Tropauer, Franz and Dilgard 1970; Burton 1975 and Boyle, di Sant'Agnese, Sack et al 1976).

The important role of the family as a medium of information - sharing is further underscored by the findings that parents are either the patient's main source (Nolan, Desmond, Herlich and Hardy 1986) or highly rated alternative source (Craft 1981; Strauss and Wellisch 1981 and Levenson, Pfefferbaum, Copeland and Silberberg 1982) of information about his disease.

Notwithstanding the proven advantages, the CF literature contains many references which attest to the shortfalls in intrafamilial communication about disease-related issues. As early as 1964, Turk depicted CF family members as being caught in a 'web of silence'. She identified communication problems within the marital relationship, between parents and siblings and between parents and the sick child. Sixty per cent of affected children had not been told their diagnosis. Shortly thereafter Meyerowitz and Kaplan (1967) claimed that attempts by affected children to learn about their prognosis, led to increased levels of stress in their sample of CF families. In similar vein, approximately one-third of parents in McCollum and Gibson's sample (1970) avoided giving their CF children any information whatsoever about their condition, despite the fact that 72% of these children raised questions about their illness. These parents feared that any communication about the disease might lead to further and more sensitive questions such as the possibility of the child's death. Kulczycki, Robinson and Berg (1969) noted two extreme cases where parents did not even allow the word 'cystic fibrosis' to be used in their homes.

Burton (1975) identified only 27% of the CF mothers in her sample as being 'good communicators' by which she meant that they recognized the importance of, as well as encouraged, open communication, while the remainder were either 'poor' or 'moderate' communicators, 40% and 33% respectively.

When communication does take place in CF families, it is usually about practical, day-to-day matters rather than discussions about the genetic or fatal nature of the disease (Turk 1964; Kulczycki, Robinson and Berg 1969; Burton 1975; Mikkelsen, Waechter and Crittenden 1978 and De Wet 1984). Many of the explanations provided by parents were vague and evasive. Respiratory symptoms were frequently referred to as 'colds' and intestinal symptoms were termed 'stomach upsets'. Parents would, in turn, rationalize that more detailed explanations would cause unnecessary distress (Tropauer, Franz and Dilgard 1970 and De Wet 1984).

Yet other studies have noted either an extreme reluctance on the part of the adolescent CF child to discuss his disease (Allan, Townley and Phelan 1974 and Bywater 1981) or a general reticence toward asking disease-related questions (Burton 1975). Nolan and co-workers (1986) noted an interesting discrepancy in the responses of CF patients and parents to a question on whether dying had ever been discussed. Whereas 64% of parents had indicated that they had discussed dying with the patients, only 44% of the patients (older than 12 years) indicated that they had spoken about dying with either their parents (n = 6) or their doctor and peers (n = 2). These responses reflected 5 discordant patient-parent responses, all being 'patient-no' and 'parent-yes'. The authors interpreted this latter finding as suggesting either guilt on the part of the parents for not having discussed dying or alternatively that 'denying' patients had responded negatively even though such discussions had taken place.

That all CF families are not necessarily overwhelmed by disease-related discussions, is illustrated by the findings of Falkman (1977) who noted that 67% of mothers felt that they could confidently answer their affected child's questions. Furthermore 42% of mothers had answered these questions with complete honesty. In a well-designed study, Phillips and his co-workers (1985) found that communicating about CF was considered to be only a 'minor' problem by 21% out of 55% of parents who had discussed the implications of the illness with their offspring. By 'minor' problem the authors meant 'limited in frequency and scope' and 'less affectively intensive' (p.183). It is nevertheless noteworthy that 22% of parents had never discussed the diagnosis with their sick child. Reasons for this failure were not presented in the study.

Most research to date has focused on communication issues between the parents and the fatally ill child. Far less is known about the information needs of siblings in these families. In fact, the present author is not aware of any published studies which have documented either the extent of knowledge or need for further information, among the siblings in CF families. Drawing from evidence in related fields, siblings want to know about the diagnosis, treatment and prognosis of the fatal illness (Koocher and O'Malley 1981), that the disease is not contagious (Johnson, Rudolf and Hartman 1979), that they are in no way to blame for the illness (Koocher and O'Malley 1981) and their own carrier status (Firth 1983 and Fitzpatrick and Barry 1986).

Several CF studies, however, report a minimum of communication about CF between parents and siblings (Turk 1964; McCollum and Gibson 1970 and Burton 1975). Parents commonly reported that siblings did not spontaneously ask about the disease which parents interpreted as their not wanting to know.

McCollum and Gibson (1970) provide a few examples of where siblings deliberately taunted an affected child with the possibility that he might die, which might well have led parents to withhold information from siblings. Similarly, Meyerowitz and Kaplan (1967) noted that levels of stress were higher in those CF families where siblings were well-informed about the illness. In contrast, Phillips, Bohannon, Gayton and Friedman (1985) report only 'minor' parental problems in communicating with siblings about disease-related concerns.

In order to ensure that siblings are kept well-informed, and in view of the difficulties inherent in such communication, it has been suggested in recent review articles that siblings might benefit from talking to health professionals (McKeever 1983) or from group discussions with other siblings in a similar position (Lobato 1983).

While the studies under review provide convincing evidence that, for many CF families, communication about the implications of CF is difficult, with one exception (Phillips, Bohannon, Gayton and Friedman 1985) these studies have not attempted to operationalize communication problems. As a result most findings are based solely on the parent's and/or researcher's interpretation of a communication problem. This has made comparison between studies difficult.

4.2.2.2 Written Information as a Source of Informational Support

Written material has become a useful adjunct to the doctor's verbal communication of information. Parents are able to study written information in their own time following which they can pose questions concerning areas of the illness about which they remain uncertain (Kupst, Dresser, Schulman and Paul 1976). Unless, however, this material is appropriate to parent and patient levels of understanding, its effects will, at best, be ineffective and at worst, may serve to heighten fears about the disease (Pless and Satterwhite 1971 and Weiner and Levitt 1984).

Reports on how CF parents view the role of written material reflect conflicting views. Burton (1975) found that most parents were generally satisfied with the written pamphlets on CF. Fathers, in particular, received the written material favourably. Burton reasoned that because fathers seldom attended clinics, they had to rely on alternative sources of information to increase their knowledge.

Other CF studies, however, report unfavourable responses on the part of parents (Lawler, Nakienly and Wright 1966; Allan, Townley and Phelan 1974 and De Wet and Cywes 1985b) and patients (Strauss and Wellisch 1981) to the published literature. Parents felt that articles emphasized the less desirable aspects of the disease such as early death and male sterility which, in turn, heightened feelings of hopelessness.

These conflicting views may well reflect the variable quality of the published literature. Having empirically evaluated 88 publications on cerebral palsy, Blasco and co-workers (1983) concluded that 58% were poorly written, out of date and misleading. It is quite conceivable that a similar evaluation of the lay CF literature might well reach similar conclusions.

In summary, while it appears that parent and patient views are divided as to the value of CF literature, were this material to be well produced, factual and not too technical and pessimistic, they may indeed welcome some written information. The argument in favour of at least a measure of supportive written material is probably best summed up by the following quotations: 'While saying too much too soon may cause unnecessary anxiety, more commonly too little is said too late, and in too ambiguous a manner' (Steinhauer, Mushin and Rae-Grant 1974) (p.837),hence '... a singular advantage of written explanatory materials is that they may be read and reread by family members if and when they are able to absorb the content'. (Van Vechten, Satterwhite and Pless 1977) (p.311). Although written materials must never replace the doctor's explanation, they can certainly supplement and reinforce it.

4.2.2.3 The Doctor as a Source of Informational Support

Several CF studies have identified the doctor as the main source of information for both the patient (Strauss and Wellisch 1981) and his parents (Allan, Townley and Phelan 1974; Burton 1975; Mikkelsen, Waechter and Crittenden 1978; Frydman 1981 and Phillips, Bohannon, Gayton and Friedman 1985). Too little information, however, remains an important source of dissatisfaction among many CF families (McCollum and Gibson 1970; Falkman 1977 and De Wet and Cywes 1985b). The finding by Falkman (1977) that 58% of her random sample of CF families had been given inadequate information is all the more noteworthy as she operationalized the concepts of 'adequate' and 'inadequate' information, the failure of which is a common flaw in many studies which assess satisfaction with doctor-patient communication (Locker and Dunt 1978).

This lack of information is most keenly felt in areas pertaining to the emotional and psychosocial aspects of managing the chronically ill child. In his study, which included a random sample of CF families, Frydman (1981) notes that 27% had received no advice regarding the sick child's overall adjustment to his illness and 38% had received no guidance regarding the child's future.

While the accumulated evidence supports the contention that parents receive too little information which is often too difficult to understand, the significance of these findings must be evaluated within the context of their methodological shortcomings. Particularly relevant is the issue of external validity, that is, the extent to which patient and parent opinions accurately reflect the care being given (Lebow 1974). It is not possible from the studies under review to know how much parents' views, on the quality and quantity of information received, are a function of actual doctor behaviour. Indeed, Falkman (1977) and De Wet and Cywes (1985b) acknowledge that their studies contain no objective measurements of how much information had, in fact, been given to their CF parents.

The subjectivity and questionable validity of these reports are further compounded by the recognition that many patients and parents, because of the distressing nature of certain information, forget much of what they are told (Burton 1975). Thus it is not possible to know conclusively whether reported dissatisfaction is justified or whether patients and parents are given adequate information which they promptly forget.

When seen within an historical context it is not difficult to understand why doctors may have failed to provide CF parents, and more particularly CF patients, with sufficient information. Indeed, an open approach towards the disclosure of information in the face of serious illness is of relatively recent origin (Reiser 1980) and continues to remain a controversial issue in medical circles (Barnard 1985 and Goldfield and Rothman 1987). This section is therefore concluded with a review of the anecdotal and empirical literature pertaining to the role of the doctor as a source of sensitive information. It begins with an historical perspective.

For many centuries doctors followed a policy of concealment, believing that the revelation of the truth about the seriousness of the patient's illness could serve only to destroy his hope. The physician's role was seen as one of comforting the patient and bad news was not considered to be part of acceptable bedside manner (Reiser 1980).

In fact, a policy of withholding the truth continued well into the first half of the twentieth century. The second half of the twentieth century, however, saw the emergence of a more empirical, though not necessarily enlightened, approach as efforts were made to measure medical and lay opinion on this issue (Aitken-Swan and Easson 1959). In a survey, in 1955, of 442 physicians regarding their attitudes towards the disclosure of sensitive information to adult patients, Fitts and Ravdin found that only 31% always or usually told patients that they had cancer. Oken in his classic study of 219 physicians found that only 12% regularly told their adult patients that they had cancer (1961). In 1965 Feifel summarized several surveys and estimated that only between 10% and 31% of doctors were in favour of telling patients that they were dying. The findings of Mount and co-workers (1974) provided still further evidence in support of a policy of concealment - only 13% of their sample of doctors felt that critically ill patients should 'always know the nature of their illness'. Only 12% believed that this information should be in the form of the 'absolute truth', while a mere 4% were of the opinion that patients 'always' wanted to know the complete truth regarding prognosis. These findings were in sharp contrast to the proportion of patients in the sample, who held these views, 78.65% and 63% respectively.

However, a study of Novack, Plumer, Smith et al in the late seventies, attested to a considerable change in attitude towards disclosure - of 264 physicians, 97% indicated a preference for telling adult cancer patients their fatal diagnosis. This trend continued into the eighties and subsequent studies conducted among medical students and physicians provide evidence in support of a policy of disclosure (Blumenfield, Levy and Kaufman 1979; Herman 1980; Greenwald and Nevitt 1982; Cohen, Ruckdeschel, Blanchard el at 1982 and Hays, Hoffman, Williams et al 1985).

A similar change in attitude towards the disclosure of information about serious illness is evident in the paediatric literature (Review articles by Share 1972; Slavin 1981 and Van Dongen-Melman and Sanders-Woudstra 1986). Advocates of the protective approach believed that children (most frequently those with cancer) and their siblings should be shielded from the knowledge of the illness and its prognosis (Agranoff and Mauer 1965; Howell 1966 and Evans 1968).

In a survey of the attitudes of 98 paediatric residents and paediatricians, Wiener (1970) found that whereas 81% believed that parents should 'always' be told the diagnosis, well over 50% of the sample believed that children should 'seldom' or 'never' be given this information. Should the child, however, request such information, 74% of residents and 56% of paediatricians would 'usually' or 'always' provide the diagnosis. Furthermore, the majority of the sample believed that the child should 'seldom' or 'never' be given an accurate description of his prognosis even if requested to do so by the child. This was in spite of the fact that a large proportion of the sample believed that the child knew or suspected the serious nature of his illness. Wiener (1970) interpreted these inconsistencies as evidence of the mixed feelings which the care of fatally ill children arouses in paediatricians.

Rea, Greenspoon and Spilka (1974) also found that 'most' paediatricians were prepared to honour parents' wishes to withhold information on prognosis from dying children. Novack and co-workers (1979) found that 18% of the sample of physicians were less likely to inform a child of a fatal diagnosis, compared to the 97% who were in favour of telling an adult patient.

The rationale behind this protective approach was threefold: firstly, many children did not seem to be overly concerned about their illness which was interpreted as a sign of not wanting to know (Richmond and Waisman 1955); secondly, it was felt that children under the age of 10 were too developmentally immature to understand the concept of death; and thirdly it was felt that the immature ego of the child was unable to cope with the fear of death (Evans 1968). Koocher (1984) summed-up this approach as follows: 'Until the 1970's a strategy of benign lies generally categorized the professional attitudes toward discussing serious illness and possible death with child patients. Presenting the difficult truth was deemed an additional burden at a time when the child was already sorely stressed' (p.577).

This approach was challenged by findings from a number of empirical studies (Vernick and Karon 1965; Waechter 1971; Spinetta, Rigler and Karon 1973; Spinetta and Maloney 1975 and Bluebond-Langer 1977) which claimed that from as young as six years old, the child was aware of the fatal nature of his illness and that any reluctance to ask questions about his illness was more likely the result of the child's perception of the adult's unwillingness to discuss the topic.

CF caretakers also advocate an open approach to information-giving (Stern 1986). Over a decade ago Pinkerton (1969) noted that 'in both adults and children, fear of the unknown represents an important source of anxiety, so that in the absence of reliable information patients and parents alike will tend to fill the vacuum with their private, horrific phantasies ... there is the need therefore to give patients the most detailed facts about the etiology, course and prognosis of the condition' (p.350). Sibinga, Friedman and Huang (1973) felt that to withhold information about sexual and reproductive issues could have a 'timebomb'-like effect on the CF adolescent, which they believed was similar to the impact of giving unrealistic or false hopes about the future and outcome of the illness.

Undoubtedly, improvements in prognoses for many chronic paediatric illnesses (Koocher 1984) have contributed to a more open approach. Slavin (1981) states that 'it is now easier for professionals to be hopeful and to offer children and parents a good dose of hope along with the whole truth about the diagnosis and prognosis' (p.5). Moreover, a recent policy statement from the American Board of Pediatrics (1987) has stated categorically that the patient and his family 'deserve to be told the truth about his or her illness' (p.829). Truth-telling, they believe, would also engender 'confidence and trust in the relationship' and would convey 'respect for the autonomy of the patient' (p.829).

While most writings in recent years advocate an open approach to communication with the seriously ill child and his siblings (Koocher 1980), Spinetta and Deasy-Spinetta (1979) caution that an open approach may not be suitable for all children and their families. Where a family is resistant to talking about the illness, these authors suggest that they be counselled to recognize the false sense of equilibrium that comes from excessive denial and the harm that such denial can cause.

Of equal concern is recent writings is how information about the diagnosis, treatment and prognosis is presented to children and their families. Accordingly, a number of articles have appeared in which the authors provide guidelines, based on their clinical and research experience, for communicating with seriously ill children and their families (Koocher 1980; Spinetta and Deasy-Spinetta 1979; Brent 1983; Myers 1983 and Greenberg, Jewett, Glick et al 1984).

For their part, Wolraich and his colleagues (1981) attribute many of the difficulties and failures in doctor-parent communication to a lack of adequate preparation to perform this task. In a survey of 24 paediatric residents, Wolraich and Reiter (1979) noted that only 41% had received any instruction in providing information, particularly in emotionally distressing situations and of these only 20% had received any form of direct criticism or feedback. This led them to comment that 'physicians would be horrified if physicians-in-training were asked to perform a lumbar puncture without having been given prior instruction and supervision, yet house officers frequently have to tell parents that their child has died or has a serious condition without having received any support, instruction or practice' (p.774).

Sack, Fritz, Krener and Sprunger (1984) in a study of 36, 3rd year paediatric residents, sought to compare their actual clinical experiences with respect to chronically ill and dying children, with the kind of educational experience which they were receiving. They found a large discrepancy between the magnitude of their clinical experience and the corresponding psychosocial curriculum pertaining to these experiences. They found 'little evidence of an organized, well-conceived format for the acquisition of knowledge and skills in this area' (p.681). They concluded their study by saying that 'helping young pediatricians find the skills and maturity to deal with the sorrow of their work remains a core task of all who would teach the care of sick children' (p.681).

To this end several attempts are now being made to impart the skills of giving stressful information at both an undergraduate (Sack 1982 and Wolraich, Albanese, Stone et al 1984) and post-graduate level (Wolraich, Albanese, Reiter-Thayer and Barratt 1981; Jewett, Greenberg, Champion et al 1982; Dickinson, Huels and Murphy 1983; Kastner, Marcuse, McGuire and Rothenberg 1985 and Desguin 1986). Hopefully, the recent policy statement by the American Board of Pediatrics (1987), making proficiency in interpersonal skills a requirement for certification, will provide further impetus to training efforts in this field.

It seems fitting to conclude with a timely warning from Reiser (1980) that the harm created by an inadequate mastery of the skills of communicating information, and more particularly bad news, may far outweigh the benefits to the patient and his family of receiving this information. Much medical education, he feels, is devoted to learning how to gather and evaluate facts, with far less time accorded to how to divulge them, 'and to divulge unskilfully and uncritically is to court inflicting grievous harm on patients' (p.841).

4.2.3 Summary

In summary, research findings concerning major sources of information used by CF families have been reviewed. The difficulties which CF families experience when it comes to talking about the fatal nature of the illness were highlighted. In spite of these difficulties, available evidence points convincingly to the positive contribution, which an open and honest approach to communication, plays in promoting overall patient, sibling and parental adjustment. Parental views concerning the value of written information were presented. While numerous CF studies have identified the doctor as the main source of informational support for CF patients and their families, many parents remain dissatisfied with the quantity and quality of this information. The role of the doctor as a source of information was reviewed within the context of changing attitudes towards the disclosure of information and the general lack of preparation to perform this task.

4.3 MEASURES

The location of the following measures in the questionnaires is detailed in Appendices 4 and 5.

4.3.1 Information Preferences

Information preferences were assessed by means of 3 Likert-type items which sought to determine how much family members wanted to know about CF. Statements were chosen both for their face validity and because similar items had been used successfully in a previous study of the information preferences of adult cancer patients (Cassileth, Zupkis, Sutton-Smith and March 1980). The 5 response options ranged from strongly agree (4) to strongly disagree (0). One of the items was negatively worded to reduce the possibility of bias due to acquiescent response set (Ware 1978). Scoring on this item was subsequently reversed. To increase the reliability of the measure, scores were summated across items to form a single Index of Information Preferences. A high score reflected a greater preference to be kept well-informed. Reliability coefficients (Cronbach Alpha) of 0.63, 0.68, 0.75 and 0.61 were obtained for fathers, mothers, patients and siblings respectively, reflecting acceptable levels of homogeneity for this Index.

4.3.2 Information Needs and Information Not Given Before

The information needs of fathers, mothers, patients and siblings on a variety of medical and psychosocial topics were measured on a 4 point rating scale. For each topic, respondents had to indicate whether they wanted 'a great deal more information', 'a little more information' or whether they had 'enough information'. A fourth category 'I do not want to know this' was included to identify any items of information which were not desired by family members. A high score reflected a greater need for information.

In addition, on each item, respondents had to indicate whether they had been given information on the topic before. A 'yes' or 'no' response was required which was scored 1 and 2 respectively. A high score indicated that a minimum of information had been provided in the past. Index measures of Information Needs and Information Not Given Before were calculated by summing responses to individual items.

Family members were also given the opportunity to list any other topics about which they might require further information.

4.3.3 Sources of Informational Support and Guidance about CF

As noted in the literature review, a family's ability to cope with chronic, life-threatening illness can be either facilitated or impeded by the availability and quality of informational support and guidance. Several measures of informational support were included in the study. These included measures of professional and lay sources of information and an evaluative measure of the doctor-parent/patient relationship as a source of information. A detailed description of these measures follows.

4.3.3.1 Frequency of Use and Perceived Helpfulness of Potential Sources of Information and Guidance about CF

Informational support was assessed by parental and patient ratings of a selection of potential sources (12 and 13 respectively) according to 'HOW OFTEN' they had obtained information and guidance from these sources over the past 12 months and according to 'HOW MUCH' helpful information and guidance they had received. The choice of sources for inclusion in the study was determined by a search of the literature and direct experience of the needs of the study sample. Sources included other parents with a CF child (other people with CF), the family doctor, doctors at the CF clinic, hospital physiotherapists, the social worker at the CF clinic, the hospital dietician, CF literature, the CF Association, prayer and faith, priest or minister, spouse (mother, father) and the local pharmacy.

Frequency of use was rated according to 3 categories (many times, occasionally and not at all) and perceived helpfulness was assessed by 4 categories (a great deal, quite a lot, not very much and none at all). By summing responses to each source of information, Index ratings of Frequency of Use and Perceived Helpfulness were obtained for fathers, mothers and patients. Acceptable levels of reliability were achieved on the former Index for fathers (alpha 0.68), mothers (alpha 0.70) and patients (alpha 0.66) and on the latter measure, alpha coefficients of 0.72, 0.71 and 0.69 were obtained for fathers, mothers and patients respectively. Where parents and patients had not found a source helpful, they were also asked to furnish reasons in their own words. Measures of both use and perceived helpfulness were included on the recommendations of Menaghan (1983) who, in a methodological review of the coping literature, had noted that the two were not inevitably associated.

Because many of the sources of information in this measure were hospital-based, it was not administered to siblings. Siblings were, however, asked to indicate how much they depended on their 'Mom' or 'Dad' for information and guidance about CF. The 5 response options were 'completely', 'very much', 'quite a lot', 'a little' and 'not at all'.

In addition, respondents had to select, from the list of potential sources, their single MOST IMPORTANT source of informational support and guidance. They were also given the opportunity to list any additional sources of CF information which they had found PARTICULARLY HELPFUL.

4.3.3.2 Doctor-Parent/Patient Communication Problems

Although effective communication about an illness and its management is an acknowledged component of paediatric care (American Board of Pediatrics 1987), it remains an area fraught with problems particularly as these relate to the exchange of information between doctors and patients (Tuckett and Williams 1984). Given that doctors are a major source of information for CF families (see literature review), an instrument was designed to assess potential problems that might arise when parents and patients tried to learn more about CF. Perceived problems in communicating with doctors were assessed, among parents and patients, by means of 8 Likert-type items the content of which was partly determined by a previously published scale of satisfaction with physician behaviour (Di Matteo and Hays 1980). To reduce response bias, items were interspersed among other measures and were worded both negatively and positively. Items were also phrased in such a way that the attribution of any failures of communication could have rested with either the doctors (e.g. 'The doctors are usually too busy to answer my questions about CF') or with the parents and patients (e.g. 'I often forget what the doctors tell me about CF').

Because of the poor reliability of single item measures (Babbie 1975), the more so in the face of the proven reluctance of respondents to criticise their doctors (Locker and Dunt 1978), an Index of Communication Problems was developed by summating scores on each of the items. Reasonably high reliability coefficients were obtained on the Index scores of fathers (alpha 0.75), mothers (alpha 0.70) and patients (alpha 0.78). Siblings were excluded from this assessment because of the low frequency of their attendance at the CF clinics.

4.3.4 Measures of Adjustment

Adjustment was assessed by means of standardized measures of Anxiety, Depression, Family Functioning and Dispositional Optimism. Several measures, both positive and negative, were included on the recommendations of previous researchers (Watson and Kendall 1983 and Byrne and Cunningham 1985) who believed that the exploratory nature of this area of research necessitated a multidimensional assessment of coping outcomes. Because these scales were not specifically designed for use among chronically ill samples, reliability data were calculated for each self-report measure and convergent validity was confirmed via tables of intercorrelations between the sets of measures. A description of the measures and their psychometric properties follows.

4.3.4.1 Measurement of Anxiety and Depression in Parents

The Hospital Anxiety and Depression Scale (HAD Scale) was used to measure anxiety and depression in fathers and mothers. The HAD scale was designed to detect the presence of anxiety and depression in general out-patient departments (Zigmond and Snaith 1983). Subsequent reports have shown that it can be used with equal reliability in community settings (Goldberg 1985). This scale was particularly suitable for use in the present study because the items did not imply the presence of psychopathology and measures of both anxiety and depression were combined in one, short scale.

The HAD scale consists of 14 items (7 anxiety and 7 depression) which are rated on a 4 point continuum. The presence of anxiety or depression is determined by summing the ratings for each scale. Scores can range from 0 to 21, with those above 10/11 being highly suggestive of the presence of anxiety or depression. The instrument has acceptable psychometric properties (Snaith and Taylor 1985 and Aylard, Gooding, McKenna and Snaith 1987). Reliability coefficients (Cronbach Alpha) of 0.82 and 0.74 were obtained for the combined scores of parents on the Anxiety and Depression scales respectively.

4.3.4.2 Measurement of Anxiety and Depression in Patients and Siblings

Two separate scales were used to assess anxiety and depression in patients and siblings:

Anxiety

Anxiety was assessed by means of the Revised Children's Manifest Anxiety Scale (RCMAS) (Reynolds and Richmond 1978). The scale comprises 28 items which reflect the presence or absence of anxiety. A total anxiety score is obtained by adding up the positive responses. In addition to a general anxiety factor, the scale is composed of 3 factor-analyzed dimensions of anxiety: physiological anxiety, worry and oversensitivity and concentration anxiety (Reynolds and Paget 1981). Reliability and validity studies have supported the usefulness of the RCMAS as a measure of chronic anxiety (Reynolds 1980, 1981 and 1985) and extensive normative data have been published on American children (Reynolds and Paget 1983). In order to reduce the length of the scale, the 9 items from the Lie sub-scale were excluded from the present study. Because this sub-scale contained no items measuring anxiety, its exclusion was unlikely to have affected overall ratings of anxiety. In addition, the wording on 7 items was slightly modified to make them more comprehensible in the South African context. The words in brackets refer to the original scale, while the underlined words reflect the modified version:

No 7 I get angry (mad) easily.

No 13 I often have a frightened feeling like 'butterflies' in my tummy (Often I feel sick in my stomach).

No 18 Other children/people (children) are happier than I am.

No 20 My feelings get hurt easily when I'm moaned (fussed) at.

No 24 It is hard for me to keep my mind on my work/school work (school work).

No 25 I can't sit still for long (I wriggle in my seat a lot).

No 28 I often worry about something awful (bad) happening to me.

Cronbach alphas of 0.79 and 0.81 were obtained respectively for patients and siblings on this Scale. These compare favourably with the published data (Reynolds and Paget 1983).

Depression

The Depression Self Rating Scale (DSRS) (Birleson 1981) was used to measure depression in patients and siblings. The DSRS consists of 18 items which were designed to measure moderate to severe depression in childhood. Items are rated for severity on a 3 point scale. A depression score is obtained by summing the responses (possible range 0-36). A cut-off score of 16 (Asarnow and Carlson 1985) was chosen as evidence of depression in the present study. The scale has been shown to be both reliable and valid (Asarnow and Carlson 1985 and Birleson, Hudson, Buchanan and Wolff 1987). Reliability ratings of alpha 0.82 and alpha 0.81 were obtained for patients and siblings respectively.

It is necessary, at this point, to stress that the diagnosis of anxiety and depression in chronically ill children is complicated by the similarity between the somatic symptoms of physical illness and those of anxiety and depression (Kashani and Hakami 1982; Fitzpatrick, Barry and Garvey 1986 and Kaplan, Grossman, Landa et al 1986). It is therefore possible for physically ill children who are not significantly depressed or anxious to score highly on these scales. To control for this potential problem in the present study, the responses of matched pairs of patients and siblings were compared (McNemar chi-squares) on ambiguous items, the results of which are presented in Appendix 14. With the exception of one item on the RCMAS ('My hands often feel sweaty'), where significantly fewer siblings agreed with the statement (30.77% vs 76.92%, Chi^2 4.5, $p = 0.0339$) no other differences were significant. Thus it seems fair to conclude that these scales were not reflecting increased levels of somatic symptomatology in the CF patients.

4.3.4.3 Measurement of Family Functioning in Parents, Patients and Siblings

The Family APGAR (Smilkstein 1978) was selected as a measure of global family functioning. The Family APGAR provides an estimate of a family's coping capabilities as seen through the eyes of individual family members. It comprises 5 items designed to assess 5 components of family functioning: adaptation (coping), partnership (role complementarity), growth (identity development), affection (emotional understanding) and resolve (commitment). Family functioning is determined by adding responses (2,1,0) to the 5 items (range of 0-10). Higher scores reflect greater satisfaction with family functioning. Scores above 7 reflect highly functional families. The reliability and validity of the measure have been thoroughly investigated among clinical and non-clinical samples (Del Vecchio, Smilkstein, Good et al 1979 and Smilkstein, Ashworth and Montano 1982). Research also supports the use of the Index among children from as young as 10 years old (Del Vecchio et al 1979). Reliability analysis yielded Cronbach's alphas of 0.79, 0.73 and 0.71 for parents, patients and siblings respectively. These levels were only marginally lower than those reported by Smilkstein et al 1982.

4.3.4.4 Measurement of Dispositional Optimism in Parents, Patients and Siblings

Dispositional Optimism, defined in terms of generalized outcome expectancies, was measured by the Life Orientation Test (LOT) (Scheier and Carver 1985). The LOT comprises 8 items which are rated on a 5 point scale (strongly agree = 4 to strongly disagree = 0). Higher scores represent higher levels of optimism. The 4 filler items, used to disguise the nature of the Test, were excluded from the present study because the 8 items were already interspersed among other measures in the questionnaires. Reliability and validity data have been obtained from a number of studies (Scheier and Carver 1985). Importantly, this measure was also independent of the effects of social desirability. Normative data are, however, limited to American college undergraduates. Reliability coefficients for parents (alpha 0.73), patients (alpha 0.85) and siblings (alpha 0.70) compared favourably with previous data (Scheier and Carver 1985).

4.3.4.5 Convergent Validity of the Standardized Scales

The convergent validity of the standardized measures used in the study was confirmed by the strength and direction of their intercorrelations (Tables 4.1 and 4.2). As shown in Tables 4.1 and 4.2, the scales which assessed positive aspects of adjustment (Family APGAR and Optimism) were negatively correlated with scales designed to measure psychopathology (Anxiety and Depression). The relatively high correlations between the measures of Depression and Anxiety, particularly in

TABLE 4.1

INTERCORRELATIONS (SPEARMAN RANK ORDER COEFFICIENTS) BETWEEN STANDARDIZED SCALES FOR PARENTS[1]

	ANX	DEP	APGAR	OPT
Anxiety	1.00	0.54***	-0.15	-0.30*
Depression		1.00	-0.33**	-0.34**
Family Apgar			1.00	0.13
Optimism				1.00

[1] n = 114
* $p < 0.001$ ** $p < 0.0003$ *** $p < 0.0001$

TABLE 4.2

INTERCORRELATIONS (SPEARMAN RANK ORDER COEFFICIENTS) BETWEEN STANDARDIZED SCALES FOR PATIENTS AND SIBLINGS[1,2]

	ANX	DEP	APGAR	OPT
Anxiety	1.00	0.68**	-0.04	-0.45
Depression	0.59***	1.00	-0.36	-0.58*
Family Apgar	-0.26	-0.30	1.00	0.41
Optimism	-0.26	-0.30	0.14	1.00

[1] n = 18 patients n = 29 siblings
[2] Findings for patients are presented in the upper triangle of the Table
* $p < 0,02$ ** $p = 0.0002$ *** $p = 0.0008$

the case of patients and siblings, support previous findings (Dobson 1985 and Wolfe, Finch, Saylor et al 1987) which have called into question the existence of anxiety and depression as separate constructs. A more in depth discussion of the psychometric properties of anxiety and depression scales is, however, beyond the scope of the present study.

4.4 RESULTS

4.4.1 Information Preferences

Family members' responses to the Index of Information Preferences were markedly skewed in favour of being kept fully informed about the illness (Table 4.3). Mean scores for all family groupings were well above the mid-point (6) of the Index.

TABLE 4.3

SUMMARY DATA ON THE INDEX OF INFORMATION PREFERENCES ACCORDING TO FAMILY GROUPING

	n	MEAN	SD	RANGE[1]
Fathers	54	10.69	1.26	7 - 12
Mothers	60	10.50	1.65	5 - 12
Patients	18	9.94	2.29	5 - 12
Siblings	28	9.00	1.92	5 - 12

1 Possible range was 0 - 12, with a higher score reflecting a greater desire for information

The % distribution of family members' responses to the individual items on the Index are presented in Table 4.4. The majority of respondents wanted as much information as possible about CF, notwithstanding the nature of this information. Only 1 mother, 1 patient and 2 siblings disagreed with this statement.

This information was, furthermore, considered to be their right by approximately 98% of parents, 90% of patients and 80% of siblings. Although a small proportion of respondents acknowledged that being fully informed about CF would only make them miserable, considerably more respondents disagreed with this statement.

Information Preferences were also examined in relation to selected sociodemographic and medical variables. None of these relationships were, however, statistically significant (p values: Appendix 15). Gender was not significantly associated with the Information Preferences of any family grouping.

4.4.2 Information Needs and Information Not Given Before

The information needs of parents, patients and siblings were analyzed in terms of individual item responses (% distribution) and according to an index measure calculated by summing responses across all items. Whereas the former analyses indicated how much family members wanted to know about specific topics, the latter score provided a total measure of their information needs. Because of the extremely low response rate to the category 'I do not want to know this', these responses

TABLE 4.4
INFORMATION PREFERENCES (% DISTRIBUTION) ACCORDING TO FAMILY GROUPING[1]

		STRONGLY AGREE	AGREE	NEITHER AGREE OR DISAGREE	DISAGREE	STRONGLY DISAGREE	FAMILY GROUPING
1	It is my right to know everything about my condition[2]	85.19	12.96	-	1.85	-	Fathers
		85.00	13.33	1.67	-	-	Mothers
		77.78	11.11	5.56	5.56	-	Patients
		46.43	35.71	17.76	-	-	Siblings
2	Knowing all the facts about CF will only make me miserable	-	5.56	12.96	51.85	29.62	Fathers
		1.67	8.33	20.00	31.67	-	Mothers
		-	16.67	5.56	44.44	33.33	Patients
		-	10.71	21.43	42.86	25.00	Siblings
3	I want as much information as possible about CF - good or bad	81.48	18.52	-	-	-	Fathers
		76.67	20.00	-	3.33	-	Mothers
		61.11	22.22	11.11	5.56	-	Patients
		25.00	46.43	21.43	7.14	-	Siblings

[1] n = 54 fathers n = 60 mothers n = 18 patients n = 28 siblings

[2] Wording altered according to family grouping

were combined with the category 'I have enough information'. This resulted in a 3 point rating scale (3,2 and 1). Similar analyses (i.e. % distribution according to individual items and an index score) were performed on responses indicating whether or not information had been provided previously. Content analyses were performed on the open-ended responses concerning any additional information needs.

Fathers and Mothers

Complete summary data on the Index ratings of Information Needs for all family members are presented in Appendix 16. Similar data in respect of Information Not Given Before are presented in Appendix 17.

On an Index with a possible range of 20 to 60, fathers and mothers obtained mean scores of 44.68 and 42.40 respectively. These scores fell slightly above the mid-point (40) of the Index, reflecting moderately high information needs. With a possible range of 20 to 40, on the Index measure of Information Not Been Given Before, mean scores of 29.19 and 28.75 were recorded respectively for fathers and mothers. These scores were only marginally below the mid-point (30) of the Index, suggesting that parents had not been given information on a substantial number of items.

The Index and specific item responses of fathers and mothers were compared by means of the Wilcoxon Sign Rank Test and the McNemar Test of Symmetry respectively. (For the purpose of McNemar chi-square analyses, individual items were dichotomized as follows: a great deal more information/a little more information, enough information). Differences between fathers and mothers on both Index ratings were non-significant. Statistically significant differences on specific items are presented in the following paragraphs.

The % distribution of fathers and mothers who wanted "a great deal more' information according to specific item content and who had 'not been given' this information before is presented in Table 4.5. (Complete % distributions, for each response category, of the specific information needs of fathers and mothers are presented in Appendices 18 and 19 respectively). The findings indicated that, in general, higher proportions of parents wanted a great deal more information about psychosocial issues and that they were less likely to have received this information.

Items for which less than 30% of fathers and mothers wanted a great deal more information included how to perform physiotherapy, when and how to take pancreatic enzymes and how CF is inherited. Significantly more fathers than mothers wanted a great deal more information on the latter two items (24.07% versus 5.56%, $\text{Chi}^2 = 8.33$, $p = 0.0039$ and 14.81% versus 27.78%, $\text{Chi}^2 = 4.46$, $p = 0.0348$ respectively). Compared to mothers, significantly more fathers also wanted a great deal more information about the symptoms of CF (22.22% versus 42.59%, $\text{Chi}^2 = 9.31$, $p = 0.0023$). Correspondingly low percentages of parents had not been given information on these items. Less than 40% of both fathers and mothers wanted a great deal more information

TABLE 4.5

TOPICS PERCEIVED AS REQUIRING A GREAT DEAL MORE INFORMATION BY FATHERS AND MOTHERS AND FOR WHICH NO PREVIOUS INFORMATION HAD BEEN GIVEN (% DISTRIBUTION)[1]

	TOPICS	FATHERS		MOTHERS	
		A GREAT DEAL MORE INFORMATION	NOT BEEN GIVEN BEFORE	A GREAT DEAL MORE INFORMATION	NOT BEEN GIVEN BEFORE
1	How the human body works	30.19	37.74	33.33	31.67
2	Symptoms of CF	42.59	20.75	26.67	15.00
3	How CF is inherited	27.78	22.64	20.00	15.00
4	When and how to administer antibiotics	39.62	39.62	36.67	30.00
5	When and how to take pancreatic enzymes	24.04	22.64	10.00	13.33
6	How to perform physiotherapy	26.42	22.64	16.67	10.00
7	How to use a nebulizer	35.85	56.60	38.33	48.33
8	Forms of exercise and sport that are suitable for people with CF	50.94	49.06	33.33	46.67
9	Healthy meals for CF children	48.15	37.74	35.00	26.67
10	New information about CF from other places in the world	79.25	54.72	68.33	56.67
11	Detailed results of any tests performed on your child when he/she attends clinic	51.85	43.40	36.67	31.67
12	Possible complications of CF	67.92	45.28	56.67	35.00
13	How to deal with your child's feelings about having CF	56.60	62.26	63.33	73.33
14	What to tell your child about his/her illness	43.40	54.72	43.33	55.00
15	What to tell your other children about CF	47.17	60.38	40.00	70.00
16	How CF might affect your child's career, social life and marriage	67.92	69.81	60.00	76.67

Continued on next page

Table 4.5 continued

	TOPICS	FATHERS		MOTHERS	
		A GREAT DEAL MORE INFORMATION	NOT BEEN GIVEN BEFORE	A GREAT DEAL MORE INFORMATION	NOT BEEN GIVEN BEFORE
17	How CF might affect your child's chances of having his/her own children	64.15	58.49	63.33	68.33
18	How serious your child's illness is	57.41	43.30	43.33	33.33
19	What to expect if your child's illness gets worse	60.38	52.83	51.67	61.67
20	How to answer your child's questions about whether he might die from his illness	50.94	64.15	61.67	76.67

1 n = 53 fathers n = 60 mothers

about how the body works, when and how to take antibiotics and how to use a nebulizer.

Whereas less than 40% of mothers wanted a great deal more information about forms of exercise and sport that are suitable for persons with CF, healthy meals for CF children and detailed results of special investigations, significantly higher proportions of fathers were in need of such information (27.78% versus 50.94%, $Chi^2 = 8.00$, $p = 0.0047$; 31.48% versus 48.15%, $Chi^2 = 4.77$, $p = 0.0290$ and 31.19% versus 51.85%, $Chi^2 = 4.77$, $p = 0.0290$ respectively). Significantly more fathers than mothers had not been given information before on the following items: How CF is inherited (22.64% versus 11.11%, $Chi^2 = 4.50$, $p = 0.0339$), how to perform physiotherapy (22.64% versus 9.26%, $Chi^2 = 5.40$, $p = 0.0196$) and detailed results of tests performed at the clinic (43.4% versus 27.78%, $Chi^2 = 4.60$, $p = 0.0325$).

At least 40% of both parents wanted a great deal more information on 10 out of the 20 topics (Table 4.6), the content of which reflected a pressing need to know much more about the emotional and psychosocial aspects of the illness. Responses, furthermore, suggested that for a large proportion of parents this information had not been supplied previously. Approximately 45% of parents wanted much more information about how they should tell the patient and his siblings about CF, while an even higher proportion of mothers (61.67%) and fathers (50.94%) needed guidance about how best to answer the patient's questions as to whether he might die from his illness.

TABLE 4.6

TOPICS ABOUT WHICH AT LEAST 40% OF FATHERS AND MOTHERS REQUIRED

A GREAT DEAL MORE INFORMATION[1]

TOPICS	ITEM NO[2]	FATHERS	MOTHERS
New information about CF from other places in the world	10	79.25 (54.72)[3]	68.33 (56.67)
How to deal with your child's feelings about having CF	13	56.60 (62.26)	63.33 (73.33)
How CF might affect your child's chances of having his/her own children	17	64.15 (58.49)	63.33 (68.33)
How to answer your child's questions about whether he might die from his illness	20	50.94 (64.15)	61.67 (76.67)
How CF might affect your child's career, social life and marriage	16	67.92 (69.81)	60.00 (76.67)
Possible complications of CF	12	67.92 (45.28)	56.67 (35.00)
What to expect if your child's illness gets worse	19	60.38 (52.83)	51.67 (61.67)
How serious your child's illness is	18	58.49 (43.40)	43.33 (33.33)
What to tell your child about his/her illness	14	43.4 (54.72)	43.33 (55.00)
What to tell your other children about CF	15	47.17 (60.38)	40.00 (70.00)

[1] n = 53 fathers n = 60 mothers

[2] Item numbers as these appear in the original questionnaires

[3] Percentages in brackets refer to proportions of fathers and mothers who had not been given this information before

Similarly high proportions of mothers (63.33%) and fathers (56.6%) needed much more guidance on how to cope with their child's feelings about having CF. Parents also wanted to know a great deal more about the future. Significantly more fathers than mothers wanted much more information about how serious the patient's illness is (57.41% versus 38.89%, $Chi^2 = 5.00$, $p = 0.0253$), while slightly more fathers than mothers also wanted a great deal more information about the complications of CF and what to expect if the illness gets worse. Over 60% of fathers and mothers were in need of much more information about issues relating to the CF patient's career, social life, marriage and reproductive prospects. New information, about CF, from other places in the world was the most sought after item of information by fathers (79.25%) and mothers (68.33%) alike, yet approximately half the sample had not received such information before.

Slightly less than 40% of fathers and mothers responded to the open-ended question concerning additional information needs. The highly individualized nature of these responses reflected a wide variety of needs which did not lend themselves to explicit categorization. Many of the parents re-iterated their desire for further information about current CF research particularly concerning newer forms of treatment and progress towards a cure. Many parents also wanted to know if some CF children were less severely affected by their illness than others. A third area about which a minority of parents wanted more information related to ways to increase the CF child's appetite. Indeed, the tone of these comments reflected a measure of distinct desperation in this regard. Additional areas mentioned included, inter alia, the effects on the child of having a chronic or terminal illness, the effects of hospitalization, how to tell grandparents and close relations about the illness, insurance issues, the effects of climate on the illness and the advisability of using alternative medicine.

The Index Information Needs of parents were examined in relation to selected sociodemographic and medical variables. (Because the difference between the Index Information Needs of mothers and fathers was not statistically significant, their scores were combined for these analyses). Only social class ($F = 5.08$, df 4, $p = 0.0009$) was significantly associated with the Index Information Needs of parents (p values: Appendix 20). Subsequent pairwise comparisons revealed significantly higher needs for social classes V (mean 50.5), IV (mean 45.29) and III (mean 47.29) compared to those of social classes II (mean 39.44), and I (mean 38.70) ($p < 0.05$). Social class accounted for 15.84% of the total variance in parental Index ratings of their Information Needs. The effect of social class was also examined separately for fathers and mothers. A significant social class effect was noted for fathers ($F = 3.20$, df 4, $p = 0.0208$) but not for mothers ($F = 2.43$, df 4, $p = 0.0588$). Further pairwise testing failed to reveal any significant differences between the mean Information Needs of fathers according to social class.

The <u>specific</u> information needs of parents (combined) were further classified according to age and Shwachman Score. The reasons for singling out these two variables for further analysis were two-fold. Firstly, empirical research has shown that specific information needs may be linked to developmental and life cycle changes (Wikler 1981) and secondly, the severity of a patient's condition may determine the nature of information given to the family (Wiener 1970). Because of the large number of statistical tests (ANOVAS) completed, a conservative alpha level (0.005) was chosen for these analyses. With the exception of a significant association between age and How to use a nebulizer ($F = 7.96$, df 2, $p = 0.0006$), neither age nor Shwachman Score were associated with the specific information needs of parents. Subsequent pairwise testing found significant differences between categories 3 and 1 ($p < 0.05$). This indicated that parents with a younger child (mean 6.69 years) required a significantly 'great deal more information' on this item than parents with an older child (mean 10.86 years) who had 'enough information'. It is worth noting that a similar, but less significant, finding was noted between this same item and Shwachman Score ($F = 4.88$, df 2, $p = 0.0095$). The significant difference between categories 3 and 1 indicated that parents with a child with a higher Shwachman Score (mean 77.95) wanted 'a great deal more information' on How to use a nebulizer compared to parents with a child with a lower score (mean 66.18) who had 'enough information' on this topic ($p < 0.05$).

Parents' Index ratings of Information Not Given Before were examined in relation to selected sociodemographic and medical variables. With the exception of a highly significant association with social class (F = 7.79, df 4, $p = 0.0001$) no other relationships were significant (p values: Appendix 20).

Subsequent pairwise testing revealed that parents from social classes V (mean 33.83), IV (mean 30.83) and III (mean 30.69) were significantly less likely to have been given information than those from social classes II (mean 26.65) and I (mean 25.38) ($p < 0.05$). Separate analyses, however, again revealed a significant social class effect for fathers (F = 7.77, df 4, $p = 0.0001$) but not for mothers (F = 1.79, df 4, $p = 0.1443$). Fathers from social classes I (mean 23.6) and II (mean 25.97) were significantly more likely to have been given information than fathers from social classes III (mean 31.7) , IV (mean 31.5) and V (mean 36.0) ($p < 0.05$). The proportion of variance in parents' and fathers' Index ratings of Information Not Given Before determined by social class was 22.38% and 39.29% respectively.

Whether or not information had been provided on specific items was also examined according to age and Shwachman Score. Again the only significant associations were between How to use a nebulizer and age (F = 18.7, $p = 0.0001$) and Shwachman Score (F = 21.00, $p = 0.0001$). The findings indicated that significantly more parents with an older CF child (mean 10.33 versus mean 6.68 years) and with a child who was more severely affected by the disease (mean 59.26 versus 69.75 Shwachman Scores) had been given this information before.

Information Needs of Patients

Summary data of patients' Index ratings of Information Needs and Information Not Given Before are presented in Appendices 16 and 17 respectively. In general, patients' total information needs were not as high as those of parents with their mean Index score (31.94) being well below the mid-point (38) of the possible range of scores. Their mean score (27.22) on the Index measure of Information Not Given Before was, however, only slightly below the mid-point (28.5) of the range. Together these findings suggest that even though patients had not been given information on numerous items they did not necessarily feel a need for very much more information.

A complete summary (% distribution) of patients' specific information needs is presented in Table 4.7. Less than 20% of patients wanted a great deal more information on 5 of the items. Only one (5.56%) patient wanted a great deal more information on how to perform physiotherapy and when and how to take pancreatic enzymes, 11.11% on how to talk to parents about their illness, with a further 16.67% requiring a great deal more information about the symptoms of CF and how to deal with feelings about having CF. Whereas most patients had been given information about physiotherapy (100%), pancreatic enzymes (94.44%) and the symptoms of CF (61.11%), only 38.89% and 33.33% respectively had been given information on how to deal with feelings about having CF and about communicating with parents about the illness. With respect to these latter two items, it is noteworthy that a large proportion of patients had neither been given this information nor did they desire further information.

TABLE 4.7

CF PATIENTS' NEEDS FOR FURTHER INFORMATION (% DISTRIBUTION) ON SELECTED MEDICAL AND PSYCHOSOCIAL TOPICS AND PERCENTAGE NOT HAVING RECEIVED THIS INFORMATION BEFORE[1]

	TOPICS	A GREAT DEAL MORE INFORMATION	A LITTLE MORE INFORMATION	I HAVE ENOUGH INFORMATION	INFORMATION NOT BEEN GIVEN BEFORE
1	How the human body works	22.22	27.78	50.00	16.67
2	Symptoms of CF	16.67	27.78	55.56	38.89
3	How CF is inherited	27.78	27.78	44.44	44.44
4	When and how to administer antibiotics	-	22.22	77.78	5.56
5	When and how to take pancreatic enzymes	5.56	5.56	88.89	5.56
6	How to perform physiotherapy	5.56	22.22	72.22	-
7	How to use a nebulizer	27.78	11.11	61.11	27.78
8	Forms of exercise and sport that are suitable for people with CF	22.22	38.89	38.89	38.89
9	What kinds of food to eat to increase your weight	44.44	11.11	44.44	38.89
10	New information about CF from other places in the world	61.11	22.22	16.67	83.33
11	Detailed results of your X rays, sputum tests etc	22.22	38.89	38.89	44.44
12	Possible complications of CF	27.78	38.89	33.33	44.44
13	How to deal with your feelings about having CF	16.67	22.22	61.11	61.11
14	What to tell friends about your illness	27.78	22.22	50.00	61.11
15	How to talk to your parents about your illness	11.11	27.78	61.11	66.67

Continued on next page

Table 4.7 continued

	TOPICS	A GREAT DEAL MORE INFORMATION	A LITTLE MORE INFORMATION	I HAVE ENOUGH INFORMATION	INFORMATION NOT BEEN GIVEN BEFORE
16	How CF might affect your career, social life and marriage	55.56	22.22	22.22	72.22
17	How serious your illness is	33.33	33.33	33.33	38.89
18	What to expect if your illness gets worse	44.44	22.22	33.33	66.67
19	How other people with CF are coping with their illness	22.22	33.33	44.45	61.11

1 n = 18

On a further 7 items, less than 30% of patients wanted a great deal more information. These included information about the human body, suitable exercise and sport for the CF person, detailed results of sputum tests, how other patients cope with CF (22.22% respectively on each item), inheritance of CF, how to use a nebulizer, possible complications of CF and how to discuss the illness with friends (27.78% respectively on each item). Approximately 45% of patients had not been given information on how CF is inherited and possible complications of the illness. One-third of the patients wanted much more information on how serious their illness is.

Items for which over 40% of patients wanted a great deal more information included the following: what kinds of food to eat to increase your weight (44.44%) what to expect if your illness gets worse (44.44%), how CF might affect your career, social life and marriage (55.56%) and the latest information about CF from other places in the world (61.11%). The trend was for increasingly higher proportions of patients not to have been given information on these topics (38.89%, 66.67%, 72.22% and 83.33% respectively).

Only 3 patients requested further information on the open-ended question. Two female patients re-iterated their need for additional reproductive information and a third patient confirmed the need to know more about current CF research.

No sociodemographic or medical variables were significantly associated with the Index measures of patients' Information Needs and Information Not Given Before (p values: Appendix 20). The suggestion, however, of a moderate positive relationship between Shwachman Score and Information Needs (r_s 0.48, p = 0.0414) and of a negative relationship between age and Information Not Given Before (r_s -0.53, p =0.0235) require further replication among a larger sample. Gender was not significantly associated with either Index rating for patients.

The 19 specific information needs of patients and whether or not they had been provided with the information were also examined according to their age and Shwachman Scores. Neither age nor Shwachman Score were significantly associated (at the 0.005 level) with either the specific information needs of patients or with whether they had been provided with the information in the past.

Information Needs of Siblings

Summary data on the Index measures of siblings' Information Needs and Information Not Given Before are presented in Appendices 16 and 17 respectively. Their mean scores on both these measures (24.29 and 18.64 respectively) fell mid-way between the possible ranges. In contrast to patients, siblings were inclined to want more information.

A complete summary (% distribution) of siblings' specific information needs is presented in Table 4.8. Items for which the largest proportion of siblings required a great deal more

TABLE 4.8

SIBLINGS' NEEDS FOR FURTHER INFORMATION (% DISTRIBUTION) ON SELECTED MEDICAL AND PSYCHOSOCIAL TOPICS AND PERCENTAGE NOT HAVING RECEIVED THIS INFORMATION BEFORE[1]

	TOPICS	A GREAT DEAL MORE INFORMATION	A LITTLE MORE INFORMATION	I HAVE ENOUGH INFORMATION	INFORMATION NOT BEEN GIVEN BEFORE
1	How the human body works	50.00	17.86	32.14	25.00
2	Symptoms of CF e.g. coughing	32.14	32.14	35.71	50.00
3	How CF is inherited	32.14	39.29	28.57	50.00
4	Different kinds of treatment for CF	53.57	25.00	21.43	64.29
5	New information about CF from other places in the world	50.00	28.57	21.43	82.14
6	Possible complications of CF	39.29	46.43	14.28	67.86
7	How to deal with your feelings about having a brother/sister with CF	32.14	28.57	39.29	60.71
8	What to tell your friends about CF	28.57	32.14	39.28	67.86
9	How to talk to your parents about CF	25.00	39.29	35.71	53.57
10	How serious your brother/sister's illness is	39.29	25.00	35.71	25.00
11	The chances of your children having CF one day	42.86	35.71	21.42	57.14
12	What to expect if your brother/sister's illness gets worse	50.00	25.00	25.00	60.71

[1] n = 28

information included the following: different kinds of treatment for CF (53.57%), how the body works (50%), new information about CF from other places in the world (50%) and what to expect if the illness gets worse (50%). With the exception of the item on how the human body works about which three-quarters of the siblings had been given information, well over 60% of siblings had not been given information on the remaining 3 topics. Approximately 40% of siblings wanted a great deal more information about the chances of their children having CF one day, possible complications of CF and how serious the patient's illness is. Whereas 75% of siblings had been given information as to the seriousness of CF, only 32.14% had been informed about the complications of CF and only 42.86% had been advised as to their chances of producing affected offspring. Less than one-third of siblings wanted much more information about the symptoms of CF, how it is inherited, how to deal with their feelings about the affected sibling and how to discuss the illness with parents and peers. Over 50% of siblings had not been given information on these items.

Only 4 (14.29%) siblings indicated that they wanted further information on the open-ended question. These respondents wanted to know how they could help the patient get better.

The Index measures of siblings' Information Needs and Information Not Given Before were not significantly associated with any of the selected sociodemographic and medical variables (p values: Appendix 20). Gender was not significantly associated with either Index measure for siblings. The specific information needs of siblings were examined only according to their age. None of these analyses yielded significant findings at the 0.005 level.

4.4.3 Sources of Informational Support and Guidance about CF

4.4.3.1 Frequency of Use and Perceived Helpfulness of Potential Sources of Information and Guidance about CF

Parents and patients rated potential sources of informational support and guidance according to how often they had been used over the past 12 months and according to the extent of their perceived helpfulness. Mean ratings were calculated for each source of support and data were ranked according to frequency of use and perceived helpfulness. An overall index of informational support, comprising all the potential sources of information, was calculated by summing responses across the items for both frequency of contact and perceived helpfulness. Findings are presented in Tables 4.9 and 4.10 for parents and patients respectively. The percentage distribution of responses to individual items is detailed in Appendices 21 to 24.

Fathers and Mothers

Both fathers and mothers depended most often for their information upon their spouse, the clinic doctors, prayer and faith and the CF literature (Table 4.9). Fathers, however, were most likely to turn to mothers for informational support, whereas mothers most often approached the clinic doctors. The following sources had been consulted far less frequently by parents: the CF Association, other parents with a CF child, the

TABLE 4.9

PARENTAL RATINGS OF FREQUENCY OF USE AND DEGREE OF HELPFULNESS OF SOURCES OF INFORMATIONAL SUPPORT AND GUIDANCE ABOUT CF[1]

SOURCE	FREQUENCY of SUPPORT[2]				HELPFULNESS OF SUPPORT[3]			
	FATHERS		MOTHERS		FATHERS		MOTHERS	
	MEAN	RANK	MEAN	RANK	MEAN	RANK	MEAN	RANK
Wife/husband	2.65	1	2.15	4	3.31	1	2.68	4
Prayer and faith	2.30	2	2.52	2	2.87	3	3.10	2
Clinic doctors	2.24	3	2.68	1	3.00	2	3.32	1
Books, pamphlets and magazines on CF	2.06	4	2.27	3	2.56	4	2.75	3
CF Association	1.72	5	1.82	6	2.20	5	2.18	6
Other parents with a CF child	1.69	6	1.98	5	2.11	6	2.40	5
Priest or minister	1.63	7	1.68	7	1.74	8	1.93	8
Physiotherapist at the hospital	1.57	8	1.53	8	1.92	7	2.02	7
Your family doctor	1.43	9	1.53	8	1.61	9	1.60	9
Dietician at hospital	1.22	10	1.08	11	1.39	11	1.15	12
Your local pharmacy	1.22	10	1.28	10	1.20	12	1.25	11
Social worker at the CF clinic	1.17	11	1.42	9	1.41	10	1.55	10
Index of support from all contributing sources[4]	20.89	(3.63)	22.08	(4.20)	25.33	(5.93)	25.98	(5.88)

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[1] n = 54 fathers n = 60 mothers

[2] Possible range: 3 = Many times 2 = Occasionally 1 = Not at all

[3] Possible range: 4 = Great deal 2 = Quite a lot 2 = Not very much 1 = None at all

[4] A combined rating was formed by summing scores across the 12 items. The possible range of scores for Frequency and Helpfulness were 12-36 and 12-48 respectively. The numbers in brackets refer to the SD's.

priest, the family doctor, the paramedical team (physiotherapists, dietician and social worker) and the pharmacist.

Written parental responses to the open-ended question as to their single most important source of information confirmed the trends revealed in the above findings. Twenty two (44.9%) fathers chose their wives, 19 (38.77%) selected the clinic doctors, with a further 7 (14.29%) selecting overseas CF literature. Only 1 father chose his priest. Five fathers did not respond to this question. Mothers, in turn, selected as their single most important source of information, the clinic doctors (84.48%), overseas literature (10.34%), the CF Association (3.45%) and faith (1.72%). Data were missing for 2 mothers.

Parental ratings of the Degree of Helpfulness of the 12 sources of informational support revealed that the most frequently used sources were also seen to be the most helpful. This was confirmed by a highly significant correlation (r_s 0.86, p = 0.0001) between the Index ratings of Frequency of Use and Degree of Helpfulness.

Clinic doctors, one's spouse, prayer and faith and the CF literature were rated as providing the most helpful information and guidance by both fathers and mothers, although the ordering of their mean ratings was again different. Whereas mothers received the highest mean helpfulness rating from fathers, clinic doctors received the highest average rating from mothers.

Both fathers and mothers rated the CF Association and other parents with a CF child as being moderately helpful sources of information, whereas they perceived the family doctor, physiotherapists, the dietician, the social worker, the pharmacist and the priest as not providing very much helpful information at all.

Some indication as to why parents had found certain sources unhelpful was provided by their written comments. In general, parents did not think that the family doctor had enough 'specialist' knowledge to advise them about CF; moreover, many parents considered themselves to be better informed about the illness than the GP's. In a few instances, the family doctor had actually expressed a reluctance to become involved 'beyond his sphere of experience'. Several parental comments also revealed an underlying bitterness, the more so in cases where a GP had missed the diagnosis of CF altogether. As a result these families felt that GP's were ill-equipped to advise them about CF.

In similar vein, parents did not feel that the local pharmacist was suitably qualified to advise them about CF. Many parents indicated that they had been unaware that a dietician was available to counsel them on nutritional issues and hence over three-quarters of the families had had no contact whatsoever with this source of information. The most frequently expressed reason for not approaching the social worker for information and guidance was the erroneously held belief that, before they could consult the social worker, parents needed to have a legitimate 'social' problem. They seemed unaware that the social worker was particularly well-positioned to offer guidance on many emotional and 'non-medical' issues. Reasons for not finding the physiotherapists helpful included personality differences, the belief by many parents that they already knew enough about this aspect of treatment and a preference for using private physiotherapists.

A few parents indicated that they were unable to attend the evening meetings held by the CF Association. Because they believed that all CF patients were different, many parents did not find it helpful to discuss their child with other CF parents. Indeed, several parents found that where conflicting opinions were expressed by different parents more questions were raised than answered.

Patients

Like their parents, CF patients also relied on the clinic doctors and prayer and faith for much of their informational support and guidance about CF (Table 4.10). Not unexpectedly patients were almost as likely to approach their mothers for information about their illness as the clinic doctors. Moreover, almost twice as many patients had approached their mothers on many occasions for information than their fathers (72.22% versus 38.89%). Mean ratings on the remaining sources of information revealed minimal utilization with a high proportion of patients reporting no contact with their pharmacist (94.44%), the hospital dietician (83.33%), the family doctor (77.77%), the CF Association (72.22%), the clinic social worker (72.22%), other CF patients (66.67%), their priest (66.67%) and the hospital physiotherapists (50%). Almost 40% of patients had not used the CF literature as a source of information about their condition.

TABLE 4.10

PATIENTS' RATINGS OF FREQUENCY OF USE AND DEGREE OF HELPFULNESS OF SOURCES OF INFORMATIONAL SUPPORT AND GUIDANCE ABOUT CF[1]

SOURCE	FREQUENCY OF SUPPORT[2]		HELPFULNESS OF SUPPORT[3]	
	MEAN	RANK	MEAN	RANK
CF Clinic doctors	2.78	1	3.17	2
Mom	2.67	2	3.50	1
Prayer and faith	2.44	3	3.11	3
Dad	2.06	4	2.61	5
Books, pamphlets and magazines about CF	1.83	5	2.78	4
Your priest or minister	1.56	6	1.78	6
Physiotherapist at the hospital	1.56	6	1.50	8
Other people with CF	1.50	7	1.44	9
Social worker at CF clinic	1.39	8	1.33	10
CF Association	1.33	9	1.61	7
Your family doctor	1.28	10	1.50	8
Dietician at hospital	1.17	11	1.22	11
Your chemist	1.05	12	1.05	12
Index of support from all contributing sources[4]	22.61	(3.79)	26.11	(6.04)

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[1] n = 18

[2] Possible range:
3 = Many times 2 = Occasionally 1 = Not at all

[3] Possible range:
4 = Great deal 3 = Quite a lot 2 = Not very much
1 = None at all

[4] A combined rating was formed by summing scores across the 13 items. The possible range of scores for Frequency and Helpfulness were 13-39 and 12-52 respectively. The numbers in brackets refer to the SD's.

In keeping with their scaled responses, written comments revealed that equal proportions of patients had selected their mothers (44.44%) and the clinic doctors (44.44%) as their most important source of information, while the remaining 2 patients had selected their father (5.56%) and prayer and faith (5.56%).

Mothers proved to be the CF patients' most helpful source of information and guidance about their illness, followed by the clinic doctors and prayer and faith. Although patients had sought information about their illness more often from their fathers than from the CF literature, they seemed to have found the latter to be slightly more informative. In fact, one-third of patients had not received any helpful information from their fathers. The remaining sources of informational support were rated as considerably less helpful by patients. Of interest was the finding that almost two-thirds of the CF patients rated their fellow patients as being of no help whatsoever as a potential source of information.

The most frequently cited reason for these low ratings of perceived helpfulness was the infrequent or total absence of contact with these sources. Three patients felt they already had sufficient information about their illness to render such contact unnecessary.

The Frequency with which patients used the various sources of informational support was only moderately correlated with their Helpfulness rating ($r_s = 0.58$, $p = 0.0121$). The significance level was also unacceptably low.

On a single item (Appendix 2: page 21) patients had to indicate if they preferred their parents to be present or absent during clinic visits. Somewhat surprisingly, 15 (83.33%) patients wanted their parents to be present during disease-related discussions with the clinic doctors.

The Index Frequency and Helpfulness ratings of fathers, mothers and patients were also examined for their relationship to selected sociodemographic and medical variables. No relationships were statistically significant (p Values: Appendix 25).

Because previous CF research (Burton 1975) had noted an association between social class and utilization of particular sources of support, further chi-square analyses were undertaken which examined the association between social class and frequency of contact with each source of informational support. (Note: Because of the strong and highly significant correlation between parental Index ratings of Frequency of Use and Perceived Helpfulness ($r_s = 0.86$, $p = 0.0001$), only ONE of the Index ratings, viz. Frequency of Use, was used in these analyses). Small cell numbers resulted in the variables being dichotomized as follows: social classes I, II/III, IV and V and many times, occasionally/not at all. Given the small sample size and hence even smaller cell numbers, similar analyses were not possible for patients. A conservative alpha level of 0.005 was chosen to minimize the likelihood of chance findings.

Social class was significantly associated with fathers' Frequency of Use of 3 of the sources of informational support. Significantly more fathers from social classes III, IV and V had not used the CF literature (24.07% versus 5.56%, $Chi^2 = 7.87$, $p = 0.005$), the CF Association (31.48% versus 9.26%, $Chi^2 = 9.61$, $p = 0.002$) and the local pharmacy (50% versus 31.48%, $Chi^2 = 8.61$, $p = 0.003$). For mothers, social class was significantly associated only with their use of the local pharmacy. Significantly more mothers from social classes III, IV and V had not utilized this source of information (51.67% versus 28.33%, $Chi^2 = 8.91$, $p = 0.003$).

Because the amount of information given to parents and patients may be influenced by the frequency of their use of sources of information, the relationship between these two measures was examined. A significant negative correlation was found only between fathers' Index ratings of Frequency of Use and Information Not Given Before ($r_s = -0.38$, $p = 0.0041$).

As already mentioned, similar ratings of informational support were not obtained from siblings. Siblings were, however, asked to indicate how much they depended on their fathers and mothers for information about CF (Table 4.11).

Nineteen (65.52%) siblings depended 'completely' or 'very much' on their mothers for such information, while only 8 (27.58%) were similarly dependent on their fathers. Ten (34.48%) siblings did not rely at all on their fathers for information about CF.

TABLE 4.11

SIBLINGS' RESPONSES (% DISTRIBUTION) TO THE QUESTION 'HOW MUCH DO YOU DEPEND ON YOUR DAD AND MOM FOR INFORMATION AND GUIDANCE ABOUT CF?'[1]

	DAD	MOM
Completely	10.34	37.93
Very much	17.24	27.59
Quite a bit	20.69	24.14
A little	17.24	10.34
Not at all	34.48	-

[1] n = 29

4.4.3.2 Doctor-Parent/Patient Communication Problems

Perceived problems in communicating with doctors about CF were assessed among parents and patients by means of an Index of Communication Problems, consisting of 8 Likert-type items. The possible range of scores was from 0, reflecting the complete absence of communication problems, to 32 reflecting the full complement of communication difficulties. The means, SD's and ranges of scores on this Index are presented in Table 4.12.

TABLE 4.12

FREQUENCY DATA ON THE INDEX OF COMMUNICATION PROBLEMS ACCORDING TO FAMILY GROUPING

	n	MEAN	SD	RANGE[1]
Fathers	54	10.30	4.24	0 - 19
Mothers	60	10.20	4.00	3 - 22
Patients	18	10.14	3.64	5 - 20

[1] Possible range was 0 to 32, with higher scores denoting greater problems with communication

Response patterns reflected a general absence of problems with communication, with the mean scores of all groupings falling well below the mid-point (16) of the possible range of scores. Although the trend was away from communication problems, a more detailed examination of responses to individual items did reveal certain problem areas (Table 4.13).

Approximately 80% of parents and well over 90% of patients felt they could count on the doctors for honest answers about the CF child's condition, while less than 6% of respondents believed that doctors withheld information lest they fail to understand the explanations. Most parents and patients also disagreed with the statement that doctors might consider them a nuisance if they asked too many questions. While the lack of privacy at clinics was not an obstacle for approximately 60% of respondents, 20% of mothers and 15% of fathers indicated that

TABLE 4.13

PARENT AND PATIENT RESPONSES (% DISTRIBUTION) TO ITEMS RELATING TO PERCEIVED PROBLEMS IN COMMUNICATING WITH DOCTORS[1]

COMMUNICATION PROBLEMS[2]	FAMILY GROUPING	STRONGLY AGREE	AGREE	NEITHER AGREE OR DISAGREE	DISAGREE	STRONGLY DISAGREE
I can really count on the doctors for honest answers about my condition	Fathers	25.93	51.95	12.96	7.41	1.85
	Mothers	33.33	48.33	11.67	6.67	-
	Patients	22.22	72.22	5.56	-	-
The doctors will think I'm a nuisance if I ask too may questions about CF	Fathers	3.70	12.96	9.25	48.15	25.93
	Mothers	1.67	13.33	11.67	50.00	23.33
	Patients	11.11	-	11.11	77.78	-
I often forget what the doctors tell me about CF	Fathers	-	24.07	7.40	59.26	9.30
	Mothers	1.67	11.67	16.67	53.33	16.67
	Patients	5.56	11.11	16.67	61.11	5.56
The doctors don't explain things to me because they think I won't understand	Fathers	-	5.56	9.25	62.96	22.22
	Mothers	-	3.33	18.33	58.33	20.00
	Patients	5.56	-	27.78	55.56	11.11
The doctors really understand how I feel about my illness	Fathers	5.56	37.04	33.33	22.22	1.85
	Mothers	13.33	38.33	26.67	15.00	6.67
	Patients	22.22	22.22	38.89	16.67	-

Continued on next page

Table 4.14 continued

COMMUNICATION PROBLEMS[2]	FAMILY GROUPING	STRONGLY AGREE	AGREE	NEITHER AGREE OR DISAGREE	DISAGREE	STRONGLY DISAGREE
The lack of privacy at the clinic makes it difficult to ask the doctors questions	Fathers	-	14.81	24.07	48.15	12.96
	Mothers	5.00	15.00	20.00	51.67	8.33
	Patients	-	11.11	33.33	55.56	-
The doctors use too many big words when they explain things to me	Fathers	-	14.81	22.22	48.15	14.81
	Mothers	5.00	13.33	18.33	51.69	11.67
	Patients	11.11	16.67	5.56	55.56	11.11
The doctors are usually too busy to answer my questions about CF	Fathers	-	1.85	22.22	51.85	24.07
	Mothers	3.33	5.00	20.00	55.00	16.67
	Patients	-	5.56	11.11	77.78	5.56

1 n = 54 fathers n = 60 mothers n = 18 patients

2 The wording on certain statements was slightly different for parents

this was a problem for them. A further 25% of fathers, 17% of patients and 13% of mothers indicated that they often forgot what the doctors told them. A substantial minority of patients (27.7%) also felt that doctors used too many big words during their explanations. This was the case for approximately 18% of mothers and 15% of fathers.

McNemar paired analysis found that parents did not differ significantly on any of the individual items. The difference between their Index ratings of Communication Problems was also non-significant (Wilcoxon Sign Rank Test).

Parent and patient scores on the Index of Communication Problems were examined in relation to selected sociodemographic and medical variables (p values: Appendix 26). Because differences between fathers and mothers were not significant, a combined score was used for these analyses.

Only the social class of parents was significantly associated with their perception of Communication Problems ($F = 4.05$, df 4, $p = 0.0042$). Subsequent pairwise testing revealed significant differences in the mean scores of parents from social classes I and V (9.29 versus 14.33) and II and V (8.94 versus 14.33) ($p < 0.05$). Only 6 parents fell into social class V and as such this finding must be treated with extreme caution. Social class was responsible for 12.93% of the variance in parental performance on this Index. The effect of social class on Index ratings was also computed separately for fathers and mothers. A significant F value was noted for fathers only (F =

3.31, df 4, $p = 0.0178$). Pairwise testing failed to produce any significant differences between the mean scores of the different social classes for fathers.

Although the negative correlations between patients' age (r_s - 0.44, $p = 0.0109$) and education (r_s - 0.47, $p = 0.0208$) failed to reach acceptable significance levels, these findings suggested that younger and less educated patients may be experiencing difficulties when communicating with clinic doctors. These findings, however, need replication in larger samples. Male and female patients did not differ significantly on this Index.

Because communication problems can impede the exchange of information between doctors and patients (Waitzkin and Stoeckle 1972), the relationships between parents' and patients' Index ratings of Communication Problems and Information Not Given Before were examined. A significant moderate correlation between these variables was found for parents only ($r_s = 0.30$, $p = 0.0003$). Regression analysis revealed that Communication Problems had contributed to 11.24% of the total variance in parental ratings of Information Not Given Before ($p = 0.0003$).

Since much of this variance could have been shared with social class (which was significantly associated with both variables), the combined effects of Communication Problems and social class on parental ratings of Information Not Given Before were examined using multivariate analysis. Together these variables determined 25.81% of the variance ($F = 7.44$, df 5, $p = 0.0001$).

Considering that social class had independently been responsible for 22.38% of the variance in parental ratings of Information Not Given Before (refer Section 4.4.2), Communication Problems had effectively contributed to less than 4% of this variance. Given the controversy surrounding the interpretation of the unique contribution of independent (i.e. predictor) variables on a dependent variable, particularly where these variables are correlated (O'Grady 1982), the relative effects of Communication Problems and social class must be treated cautiously. Notwithstanding these theoretical precautions, these findings have further underscored the important role of social class in this study.

4.4.4 Relationships Between CF Knowledge and Selected Measures of Information

In the previous chapter the relationship between CF Knowledge and selected sociodemographic and medical variables was explored. In this section findings are presented which examined relationships between CF Knowledge and Index ratings of Information Not Given Before, Sources of Information and Doctor-Parent/Patient Communication Problems.

4.4.4.1 CF Knowledge and Information Not Given Before

Parental CF Knowledge was negatively correlated with Information Not Given Before (r_s -0.52, $p = 0.0001$). When considered separately the correlations between these two variables remained highly significant for fathers and mothers (r_s -0.53, $p = 0.0001$ and r_s -0.47, $p = 0.0001$ respectively). A moderate and statistically significant negative correlation was also found between patients' CF Knowledge and Information Not Given Before (r_s -0.63, p = 0.0041). The CF Knowledge of siblings was not significantly correlated with their Index rating of Information Not Given Before, although the direction of the correlation was negative.

Parental ratings of Information Not Given Before contributed to 27.7% of the total variance in parental CF Knowledge (p = 0.0001). Because social class was again associated with both these variables, the combined effect on parental CF Knowledge of Information Not Given Before and social class was computed. Together these variables accounted for 59.04% of the variance in parental CF Knowledge (F = 24.93, df 5, p = 0.0001). This meant an increase of 5.57% compared to that contributed by social class independently.

4.4.4.2 CF Knowledge and Sources of Informational Support

The CF Knowledge of parents and patients was examined in relation to their Index ratings of Frequency of Use of sources of information about CF. Parental CF Knowledge showed only a weak and barely significant positive correlation with Frequency of Use (r_s 0.22, p = 0.0181).

Because these global measures were unable to detect the effect of individual sources of information on CF Knowledge, further analyses (ANOVAS) examined the relationship between Frequency of Use of each source of informational support. These analyses were computed for fathers and mothers only. Since these analyses entailed a large number of calculations, a conservative alpha level was set at 0.005. The more frequent use of the CF literature ($F = 9.87$, df 2, $p = 0.0002$) and greater contact with the CF Association ($F = 9.92$, df 2, $p = 0.0002$) were significantly associated with the CF Knowledge of fathers. Subsequent pairwise testing indicated that fathers who had no contact with the CF Association or had never read the CF literature had significantly lower mean Test scores than fathers who utilized these sources many times or occasionally. Where fathers had had no contact with the CF Association they had significantly lower average Test scores (mean 41.06) than fathers with occasional (mean 50.39) or more frequent contact (mean 49.57) ($p < 0.05$). Similarly fathers who consulted the CF literature many times had a significantly higher average Test score (mean 54.67) than fathers who had only occasional (mean 47.57) or no contact (mean 43.76) with this source over the past 12 months ($p < 0.05$). Only the amount of contact with the CF Association had a significant effect on the CF Knowledge of mothers ($F = 9.64$, df 2, $p = 0.0003$). Where mothers had no contact with the CF Association they too had lower average Test scores (mean 41.78) than mothers with either occasional (mean 50.43) or more frequent contact (mean 49.5) ($p < 0.05$).

4.4.4.3 CF Knowledge and Doctor-Parent/Patient Communication Problems

Whereas the Index of Communication Problems was significantly correlated, in a negative direction, with parental (r_s -0.26, $p = 0.0057$) and fathers' (r_s -0.35, $p = 0.0037$) CF Knowledge, it was not significantly correlated with the CF Knowledge of mothers. Although the correlation between patients' CF Knowledge and their performance on the Index of Communication

Problems was moderate, the significance level was very low (r_s - 0.53, $p = 0.0231$).

Using regression analysis, parental ratings of Communication Problems were found to determine only 7.42% of the total variance in their CF Knowledge ($p = 0.0034$). When the combined effect of Communication Problems and social class was computed, Communication Problems added less than 1% to the variance already accounted for by social class independently ($F = 24.93$, df 5, $R^2 = 0.5358$, $p = 0.0001$). These findings once again confirmed the importance of social class as a significant factor influencing parental CF Knowledge.

4.4.5 Relationships Between CF Knowledge and Adjustment

In this section findings are presented which explored the relationship between CF Knowledge and coping outcomes, operationalized in terms of standardized adjustment scales. Since the literature has also suggested that the lack of knowledge about an illness can be a significant source of stress for families with a chronically ill child (Harrison 1977 and Chesler and Yoak 1984), findings which examined relationships between family members' Index ratings of Information Needs and their adjustment scores are also presented.

4.4.5.1 Personal and Family Adjustment in CF Family Members

Parents

The mean adjustment scores for parents are presented in Table 4.14. In general, findings reflected high levels of Family Functioning and Optimism in parents and a marked absence of psychopathology and the mean Anxiety and Depression scores were well below the recommended cut-off figure (10/11) indicative of clinical disorder (Zigmond and Snaith 1983). The mean Family APGAR scores for fathers and mothers also fell within the range (7-10) considered to indicate a 'highly functional' family

TABLE 4.14

SUMMARY DATA ON PARENTAL PERFORMANCE ON THE STANDARDIZED ADJUSTMENT SCALES[1]

	MEAN	SD	POSSIBLE RANGE	OBSERVED RANGE
HAD Scale[2]				
Anxiety				
Fathers	6.35	2.76	0 - 21	0 - 16
Mothers	7.35	4.64	0 - 21	1 - 18
Depression				
Fathers	3.74	2.57	0 - 21	0 - 10
Mothers	4.32	3.27	0 - 21	0 - 13
Family APGAR				
Fathers	8.22	1.83	0 - 10	2 - 10
Mothers	7.92	2.26	0 - 10	2 - 10
Optimism (LOT)				
Fathers	22.96	4.14	0 - 32	17 - 32
Mothers	22.77	4.50	0 - 32	9 - 31

1 n = 54 fathers n = 60 mothers
2 Hospital Anxiety and Depression Scale

(Smilkstein 1978). Fathers and mothers also displayed significantly more Optimism than a normative sample of male (mean 21.03) and female (mean 21.41) college students (p = 0.0084 and 0.0022 respectively) (Scheier and Carver 1985). Differences between the scores of fathers and mothers were not significant on any of the scales.

Patients and Siblings

The scores of patients and siblings on the adjustment scales are summarized in Table 4.15.

As was the case with their parents, patients and siblings showed a notable lack of psychopathology. The mean Depression scores of both groups fell well below the recommended cut-off score (16) for clinical depression (Asarnow and Carlson 1985). Mean Anxiety scores for patients and siblings were compared with normative data from a national sample of American boys and girls (Reynolds and Paget 1983). Male patients (n = 9) and siblings (n = 13) received lower mean Anxiety scores than the normative group but these differences were not significant. Differences between the mean Anxiety scores of female patients (n = 9) and siblings (n= 16) revealed significantly lower Anxiety scores than the American sample (p = 0.0434 and p = 0.0243 respectively). (A more detailed presentation of Anxiety data according to sub-scale scores are presented in Appendix 27 for patients and siblings). The mean ratings of Family Functioning for both patients and siblings fell within the highly functional range of scores. Differences between the average Optimism scores of male and female patients and siblings and

American college students were not significant. Given the small sample numbers in these analyses, these findings must be treated with caution.

TABLE 4.15

SUMMARY DATA ON THE STANDARDIZED SCALES OF ADJUSTMENT FOR PATIENTS AND SIBLINGS[1]

	MEAN	SD	POSSIBLE RANGE	OBSERVED RANGE
Depression (DSRS)				
Patients	6.94	4.98	0 - 36	0 - 19
Siblings	6.59	3.57	0 - 36	0 - 14
Anxiety (RCMAS)				
Patients	9.22	6.92	0 - 28	1 - 25
Siblings	10.10	7.50	0 - 28	0 - 25
Family APGAR				
Patients	8.06	1.63	0 - 10	5 - 10
Siblings	7.07	2.07	0 - 10	2 - 10
Optimism (LOT)				
Patients	22.50	5.58	0 - 32	14 - 32
Siblings	20.04	3.60	0 - 32	14 - 28

[1] n = 18 patients n = 29 siblings

4.4.5.2 CF Knowledge and Adjustment

The relationship between the Composite CF Knowledge of family members and their performance on the standardized adjustment scales was examined by means of Spearman correlations (Table 4.16). Because none of the differences between the scores of fathers and mothers on the adjustment scales were significant, they were combined for these analyses.

TABLE 4.16

INTERCORRELATIONS (SPEARMAN COEFFICIENTS) BETWEEN COMPOSITE CF KNOWLEDGE AND PERSONAL AND FAMILY ADJUSTMENT ACCORDING TO FAMILY GROUPING[1]

	COMPOSITE CF KNOWLEDGE		
	PARENTS	PATIENTS	SIBLINGS
Depression	-0.33^{***}	-0.22	-0.09
Anxiety	-0.04	-0.30	-0.04
Family APGAR	0.22^{**}	0.26	0.20
Optimism	0.13^{**}	0.54^{*}	0.19^{*}

1 n = 114 parents n = 18 patients n = 29 siblings

* $p < 0.02$ ** $p < 0.01$ *** $p < 0.0003$

The low negative correlation between parental CF Knowledge and Depression was the only finding to reach an acceptable level of significance. The positive relationships between parental CF Knowledge and Family Functioning and between family members' CF Knowledge and Optimism while suggesting that a relationship might exist, require replication. Although not all findings were significant, the direction of the correlations between CF

Knowledge and Anxiety and Depression were negative for all family groupings.

Given the discrepant nature of the findings between family members' ratings of the CF patient's Present and Future Health and their open-ended responses concerning their fears for the patient's future (Section 3.3.3), the author decided not to examine these ratings in relation to personal and family adjustment. Such analysis was further contraindicated by the small cell frequencies in many of the response options on these two items. Notwithstanding the failure of the present study to reliably assess family members' perceptions of the patient's health, the effect of such knowledge on adjustment remains an important focus for future research, particularly in view of the life-threatening nature of this illness with its potential to affect coping.

4.4.5.3 Information Needs and Adjustment

Relationships between Index ratings of Information Needs and personal and family adjustment are presented in Table 4.17 for all family groupings.

Parents who had higher Information Needs also reported significantly higher levels of Depression and Anxiety. The correlation with Depression was, however, slightly stronger and more highly significant. The relationships between patients' and siblings' Information Needs and their self-reported Depression and Anxiety were barely significant. Only parental Optimism showed a weak negative relationship with their

Information Needs. The significance of this correlation was, however, low. Information Needs were not significantly associated with Family Functioning.

TABLE 4.17

RELATIONSHIPS (SPEARMAN CORRELATION COEFFICIENTS) BETWEEN INDEX RATINGS OF INFORMATION NEEDS AND PERSONAL AND FAMILY ADJUSTMENT ACCORDING TO FAMILY GROUPING[1]

	INFORMATION NEEDS		
	PARENTS	PATIENTS	SIBLINGS
Depression	0.42^{****}	0.31^{*}	0.43^{*}
Anxiety	-0.21^{***}	0.57^{*}	0.48^{*}
Family APGAR	-0.13	0.03	-0.77
Optimism	-0.21^{**}	-0.33	-0.20

[1] n = 114 parents n = 18 patients n = 29 siblings

$^{*} < 0.05$ $^{**} < 0.005$ $^{***} < 0.001$ **** $p = 0.0001$

4.5 SUMMARY

In summary, this chapter has reviewed the literature concerning the role of information in facilitating a family's ability to cope with chronic illness, and the main sources used by CF families in securing information about the illness. This was followed by the measurement of family members' information preferences, their information needs, what CF information they had received in the past, their main sources of information and guidance about CF and the extent to which they experienced problems in communicating with doctors when seeking information. Important sociodemographic and medical correlates of these measures were identified. The effect of not having been given information about CF and of experiencing communication problems on current CF Knowledge was also examined. Finally both the CF Knowledge and the Information Needs of family members were examined in relation to their personal and family adjustment. Findings from Chapters 3 and 4 are discussed in the following chapter.

CHAPTER 5

DISCUSSION

5.1 INTRODUCTION

The discussion of the findings has, as far as possible, followed the order of their presentation in the thesis. As the CF knowledge of families was significantly related to such factors as prior information - giving, the presence of communication problems with doctors and sources of information, a degree of overlap between the respective sections was unavoidable. In order to minimize repetition, the discussion of significant findings concerning relationships between sociodemographic and medical variables and CF knowledge and the various 'information' measures was incorporated in a single section. Recommendations, together with the implications of selected findings for CF families' ability to cope with chronic, life-threatening illness are, whenever appropriate, woven into the general text of the discussion.

5.2 CF KNOWLEDGE

As the life expectancy of CF patients increases, parents are being given increasing responsibility for the home care of these children. In addition many adolescent and adult CF patients are assuming more independent control over their medical care. Family members, and in particular parents and patients, therefore require an adequate and detailed knowledge of the symptomatology, treatment and future management of this disease. An important aim of the present study was to assess how much family members knew about the medical aspects of CF.

Responses to the 63 multiple choice questions on the CF Knowledge Test revealed that family members were far more knowledgeable about the general characteristics of the disease, gastrointestinal symptomatology and treatment, physiotherapy and anatomy than they were about nutrition, general and specific respiratory symptomatology and treatment, genetics and terminology. Parents and patients correctly answered approximately 75% of the medical questions while siblings successfully answered approximately 65% of the items. (Overall scores increased by less than 3% on the Revised version of the CF Knowledge Test).

CF patients displayed levels of knowledge equivalent to those of their fathers and mothers as evidenced by the lack of statistically significant differences between their performances on either the Composite Test or its 9 sub-sections. Moreover, the findings that mothers and fathers scored significantly better than patients on only 4 (6.35%) and 2 (3.17%) of the items respectively, suggest a high degree of similarity with respect to the content of this knowledge . In contrast, the total scores of siblings on both the Composite and Revised Tests were significantly lower than those of fathers, mothers and patients. In addition, they were significantly less informed than mothers, fathers and patients on a meaningful proportion of the individual items (20.63%, 14.29% and 12.7% respectively). Given the small sample numbers involved in these intra-familial comparisons, these findings must be viewed with caution.

In many respects these findings confirm those of previous studies (Kulczycki, Robinson and Berg 1969; Tropauer, Franz and Dilgard 1970; Bywater 1981; Strauss and Wellisch 1981 and Nolan, Desmond, Herlich and Hardy 1986) which also reported adequate levels of medical knowledge in their CF samples. The present research, however, went beyond that of previous work. Whereas most of these latter studies, with the exception of Nolan and co-workers (1986), had failed to indicate how knowledge of CF was measured, the present research used a reliable and valid instrument. The present study also included a sample, albeit small, of siblings whose knowledge of CF has, to date, remained untested. As was the case in the Nolan et al (1986) study, parents and patients were adequately informed in certain areas of CF, while in others, such as genetics and reproductive risks, their knowledge was weak and seemingly without understanding.

By its very nature as a criterion-referenced measure, an important aim of the CF Knowledge Test was to provide an accurate description of what family members did or did not know about CF. The remainder of this discussion focuses on their performance in the various domains of knowledge.

5.2.1 General CF Knowledge, Respiratory Symptomatology and Treatment, Physiotherapy and General Medical Knowledge

According to Goodchild and Dodge (1985) 'for most patients, respiratory disease is the most serious manifestation of cystic fibrosis and the rate of its progression determines the well-being and life span of the individual' (p.53). Thus for these authors the primary goals of treatment are the 'control of infection and removal of thickened bronchial secretions' (p.53). These goals are achieved principally through the administration of antibiotics, chest physiotherapy with coughing, and exercise.

Most family members correctly answered questions relating to the presence of mucous secretions in CF children and recognized their contribution to overall morbidity. Family members were similarly well-informed regarding the important role of regular physiotherapy and exercise in the removal of these harmful secretions. It is noteworthy that almost double the amount of patients (22%), compared to parents and siblings (12% and 14% respectively), did not believe that physiotherapy was necessary when a child was well. In the presence of a chest infection, however, most parents and patients endorsed the need for daily physiotherapy. The discrepant patient responses underscore the need for health professionals to include the patients' perspective in their explanations of compliance (Conrad 1985). Whereas medically the aim of physiotherapy is to keep the lungs free of secretions, patients may have entirely different objectives. Primarily they want to be able to lead a normal life, while at the same time having to cope with a chronic illness (Smith 1986). To the extent that physiotherapy impinges on these goals of normality so will compliance be concomitantly affected, irrespective of knowledge espousing its benefits.

Even though the percentages of incorrect responses were small, it is nevertheless important that all parents and patients recognize that physiotherapy be performed in children of all ages BEFORE meals to minimize vomiting which may be associated with post-prandial postural drainage (Hubbard and Mangrum 1982).

The finding that only a small proportion of parents and patients had not been given information on physiotherapy would explain their generally high knowledge on these items. Data from the present study did not support Davids' (1986) contention that much of the non-compliance with physiotherapy among her lower class families was attributable to their lack of interest in learning more about this form of treatment. Indeed, of the mothers and fathers who wanted a great deal more information about physiotherapy, approximately 60% and 90% respectively came from social classes III, IV and V. These data, instead, provide further evidence that health professionals misinterpret the diffidence of the lower social classes as reflecting less of a desire for information (Waitzkin 1984).

Despite the uniformly high proportion (over 95%) of parents and patients who recognized the importance of exercise and sport in the management of CF, approximately 51% of fathers, 33% of mothers and 22% of patients still wanted a great deal more information about suitable forms of physical exercise for CF patients. The finding that significantly more fathers than mothers wanted a great deal more information about exercise and sport is particularly noteworthy as this knowledge would also enable fathers to become more actively involved in the management of the patient. A knowledge of sport, in particular, would create the opportunity for week-end involvement on the part of fathers, who might otherwise be prevented from more frequent participation in treatment because of the pressures of employment. In addition to the medical advantages of outdoor exercise and sport, these activities also enable the CF patient to interact normally within the community. Yet almost 21% of siblings felt that sport was contraindicated for the CF patient because it might lead to excessive coughing. Given their potentially valuable role in encouraging the CF patient to take part in outdoor activities, this fallacy needs to be removed.

Although the majority of respondents recognized that a persistent cough was not reason enough to keep the CF child from attending school, over 10% of parents and patients held the mistaken belief that the CF child should be kept at home in case he infected his classmates. Given the potential for unnecessarily disrupting the patient's school attendance, it is important that all family members know that the respiratory symptoms of CF are not infectious. When seen against the finding by De Wet (1984) that most CF children strongly resent missing school, removing this misconception becomes even more imperative.

It is, furthermore, essential that similar misconceptions be clarified in the minds of school personnel. De Wet (1984) found that ignorance among teachers about CF symptoms considerably increased maternal anxiety. In a few instances teachers regarded the need for the continuous administration of antibiotics as originating more from the over-protective attitude of the mother than as being an essential aspect of medical treatment.

The distinction between bacterial and viral infections was far from clear in the minds of family members. Although it might be argued that such a distinction is of little more than academic value to the CF family, it is nevertheless advisable that parents be made aware that CF children are no more susceptible to the common cold and other viral respiratory infections than their siblings and friends (Goodchild and Dodge 1985). Yet almost 75% of parents and siblings held this misconception.

It is also important that parents, particularly those with younger CF children, be advised that viral respiratory tract infections occur frequently in all children irrespective of their underlying health status. The more so given De Wet's (1984) finding that mothers tended to see themselves as determinants of both good and bad health in patients. Thus mothers might well ascribe the frequent incidence of viral chest infections to a treatment omission on their part. Parents should also know that antibiotics have no effect on common colds and viral chest infections or to quote Sir William Osler that 'there is just one way to treat a cold and that is with contempt' (quoted in Isaacs 1987, p.44). Over 25% of all respondents in the present study erroneously believed that antibiotics were an effective form of treatment for viral infections.

In similar vein, the knowledge that asthma and allergies can occur frequently in the CF child (information possessed by slightly more than 50% of parents and patients) might alert parents to possible alternative causes of persistent respiratory symptoms in their child, especially as these can be well controlled with appropriate medication.

Approximately one-third of fathers and patients and almost 50% of siblings mistakenly believed that cough mixtures were a valuable adjunct to the treatment of lung infections. Perhaps of even greater concern was the discovery that well over 60% of parents and 70% of patients believed that nebulized antibiotics should be given before physiotherapy. As inhaled antibiotics are less likely to penetrate airways which are obstructed by thick and sticky mucus this error could result in less effective treatment. While this gap in their knowledge is most likely explained by the finding that over 50% of parents and 30% of patients had not been given information on nebulization before, many patients failed to recognize that they required further information. Over 60% of patients felt that they had enough information on nebulization, with over 80% also indicating that they were sufficiently well-informed about when and how to administer antibiotics. Equivalent findings for parents were considerably lower. Considering that older patients will be assuming more responsibility for their treatment, it is imperative that areas of ignorance and misconception be identified and clarified by the medical team, the failure of which could lead to inadvertent non-compliance.

Without equivalent findings from other CF studies, it was not possible to compare levels of knowledge about respiratory symptoms and treatment. Nolan and co-workers (1986) tested parents and patients on only one aspect of respiratory treatment, namely the reasons for aerosol therapy, about which respondents were well-informed. The present findings did, however, confirm those of Harbord and colleagues (1987) who also noted that their CF patients were more knowledgeable about chest physiotherapy than antibiotics.

5.2.2 Gastrointestinal Symptomatology and Treatment and Nutrition

As a result of pancreatic insufficiency, most CF patients suffer from varying degrees of malabsorption which results in frequent offensive, abnormally bulky stools (Goodchild and Dodge 1985) and a significant loss of calories from unabsorbed nutrients (Dodge 1985). In addition, the increased energy demands created by the presence of chronic respiratory infection and daily physiotherapy, may compromise the nutritional status of the CF child still further. Thus the control of malabsorption, together with an increased intake of calories, have become essential aspects of the overall management of the CF patient (Francis 1987). Unlike a course of antibiotics, however, which carries an explicit set of instructions for administration, both pancreatic enzyme replacement and nutritional manipulation are almost entirely dependent on the specific requirements of the individual CF patient (Francis 1987). To this end a knowledge of the rationale and underlying principles of these aspects of management are indispensable.

Patients obtained scores of over 80% and had consistently fewer errors than parents on all items relating to pancreatic supplementation. This knowledge may well have accrued as a result of their direct 'trial-and-error' experience of controlling the unpleasant symptoms resulting from malabsorption (Boyle, di Sant'Agnese, Sack et al 1976 and Strauss and Wellisch 1981). Furthermore, only one patient indicated that he had not been given information before on when and how to take pancreatic enzymes.

An applied (i.e. working) knowledge of enzyme replacements is particularly important to the older CF patient as he begins to exert more control over his food intake. Nolan and his team (1986) noted that over one-fifth of the CF children in their sample did not know the reason for this form of treatment, while Harbord and co-workers (1987) found that approximately one-third of their older CF patients (7 - 18 years) did not understand the underlying mechanism of pancreatic supplementation. On a related question, 3 (16.67%) patients in the present study failed to connect the dosage of pancrease to the amount of fat in the diet.

Because teachers often forget or incorrectly administer pancreatic supplements (De Wet 1984), it is also important that the younger school-going CF child knows that enzymes are necessary with all meals and snacks. Two patients in the present study thought that they were only necessary with the 3 main meals, suggesting the possibility of inadvertent non-compliance. To prevent potential embarrassment and to maximize absorption, it is essential that ALL patients know exactly how, when and why they are taking pancreatic enzymes.

Assuming that there is a connection between parental (particularly maternal) knowledge of treatment and subsequent compliance (see Literature Review Section 3.2.3.1), a meaningful proportion of CF children in the present study may not have been receiving adequate levels of enzyme replacement therapy. Almost 25% of fathers and 20% of mothers either incorrectly answered or were unaware that it was necessary to take enzymes with all meals and snacks, while a further 25% and 22.22% of mothers and fathers respectively did not know the best time to administer this treatment. In addition, approximately 30% of parents were unaware that the dosage of enzymes should be tailored to the amount of fat in the meal. The finding that less than 15% of mothers and approximately 22% of fathers had not been given information on when and how to take pancreatic enzymes may partly explain these findings. It also suggests that, when provided, this information may have been forgotten or misunderstood by some parents.

Despite the potential for inadvertent non-compliance caused by an incomplete knowledge of pancreatic supplementation, over 60% of mothers and almost 90% of patients felt that they had enough information about when and how to take pancreatic enzymes. In contrast, only one-third of fathers felt that they had sufficient knowledge in this respect. This finding indicates that a significant proportion of mothers and patients may not be aware of gaps in their knowledge concerning this aspect of treatment. It is recommended that the medical team takes the initiative in identifying and supplementing mistaken or missing knowledge in patients and parents.

Siblings were, in general, reasonably well-informed about the treatment of gastrointestinal symptoms, having obtained scores which ranged from 65% to 86%, the more so given the highly individualized nature of this treatment. Although it is arguably not critical for them to have this knowledge, it is noteworthy that information about the different kinds of treatment for CF, was the most sought after item of information amongst siblings. Almost 54% of siblings required a great deal more information on this topic.

Although a minimum of dietary manipulation is advocated for CF patients (Goodchild and Dodge 1985), their heightened nutritional needs remain a major focus of treatment (Hubbard and Mangrum 1982). Research has estimated that CF patients need an energy intake of approximately 150% of the recommended daily allowance of persons of equivalent age and sex to compensate for their increased energy expenditure and nutritional losses through malabsorption (Dodge 1985). Thus an increased calorie intake becomes central to nutritional management (Francis 1987). This, in turn, presupposes that parents and patients possess at least a minimum knowledge of how the energy content of food is calculated together with some knowledge concerning the calorie content of common foodstuffs. Such basic knowledge was, however, lacking in a significant proportion of the present sample.

In general, respondents were far less aware of the energy properties of calories compared to their more popular association with weight gain. No doubt the role of the media was instrumental in this finding. Almost half the sample of parents and patients also mistakenly believed that proteins were a better source of energy than carbohydrates for the CF patient. Since protein will not be used for growth unless adequate energy intake is achieved, it becomes an 'expensive' source of energy for the CF patient (Francis 1987). Given the added financial costs to the family, were protein to be used as a major source of energy, the essential role of carbohydrates in the CF patient's diet cannot be emphasized too strongly.

Respondents, furthermore, showed considerable confusion as to whether or not fat should be included in the CF patient's diet. Given that fat is a valuable source of energy for the CF patient, the findings that approximately 50% of parents and almost 90% of patients advocated its total exclusion, raise serious doubts as to whether patients were not inadvertently being deprived of an essential source of energy. When viewed against the concern felt by many CF patients about their excessive thinness (Boyle, di Sant'Agnese, Sack et al 1976 and Strauss and Wellisch 1981) and the finding from the present study that almost 45% of patients wanted a great deal more information about what foods they should eat to increase their weight, this misconception takes on an even greater significance.

Rosenstock (1985) has pointed out that in many chronic illnesses patients and their families may have to 'unlearn' what they have already learned as new treatment regimens are shown to be better than old ones. Thus it seems that many respondents had not yet 'unlearned' the earlier treatment approach which had advocated a restricted fat intake (Passero, Remor and Salomon 1975). McCollum and Gibson (1970), however, found that even though an unrestricted diet had been encouraged in their CF clinic for almost three years, two-thirds of the parents in their sample continued to impose dietary restrictions on their affected children. Parents claimed that dietary permissiveness led to an increase in 'the frequency and foulness of stools and flatulence' which, in turn, led to '... adverse social consequences' (p.575). This finding suggests that irrespective of clinic policy regarding dietary manipulation, because of their own fears regarding a loss of control, parents may be reluctant to give the CF patient freedom of choice over his diet. (At the time of this latter study pancreatic treatment was not as effective in controlling steatorrhoea as it is today).

To be successful nutritional counselling will need to provide the 'facts' (for example, the potency of newer enzyme replacements), together with a recognition of the emotional difficulties which parents are likely to encounter as they exercise less control over the patient's diet.

The generally poor performance on the nutritional items might also be explained by the finding that parents and patients seldom approached the dietician for information. Considering that she is probably one of the most appropriate and skilled team members to provide such counselling, her underutilization is cause for concern. (This issue is addressed in Section

5.6). Although the present author is not aware of any other CF studies which have specifically assessed levels of basic and applied nutritional knowledge, a recent study of juvenile diabetic patients (Lorenz, Christensen and Pichert 1986) highlighted significant deficits in their dietary knowledge, even in children who had received direct instruction from a dietician.

Given the traditional sex role divisions in our society, it was unexpected, but nonetheless welcome, that almost 50% of fathers wanted a great deal more information about healthy meals for CF children. It is important that doctors and dieticians recognize the information needs of fathers in this regard, particularly as it could lead to their greater involvement in the patient's care.

Dietary patterns are likely to influence the whole family. Thus it was not surprising that siblings shared many of the misconceptions held by parents and patients concerning the role of fat in the diet. They too lacked a basic knowledge of calories and their relationship to weight gain and increased energy. Although not questioned specifically about their desire for more dietary information, as already noted, over 50% of siblings requested a great deal more information about aspects of CF treatment. Furthermore, in view of previous findings that siblings may, as a result of the disease, feel neglected in the family (Kerner, Harvey and Lewiston 1979 and Phillips, Bohannon, Gayton and Friedman 1985), together with their wish to help with treatment (Mikkelsen,Waechter and

Crittenden 1978), this information might well enable them to become more involved, if only on a cognitive level.

5.2.3 GENETICS

Performance on genetics items highlighted inadequacies in the knowledge of all family groupings. Patients and siblings, in particular, performed poorly on these items as evidenced by average sub-section scores of less than fifty per cent. Moreover, insofar as questions on genetics are highly interrelated, with knowledge of one aspect presupposing knowledge of closely related aspects (Leonard, Chase and Childs 1972), a large proportion of parents and children in the present sample could be described as having knowledge without understanding. Thus while a meaningful proportion of parents (83.33%) knew that the CF gene had to be carried by both parents in order to give birth to an affected child, a far smaller proportion of respondents was able to translate this knowledge into actual recurrence risks for future pregnancies.

Where probabilities of recurrence were stated as percentages rather than odds, respondents were even less successful in obtaining correct answers. Leonard, Chase and Childs (1972), in fact, suggested that a poor grasp of concepts such as probability was a main cause for CF parents' poor recall and misunderstanding of genetic information. The finding that over 45% of parents gave themselves a mere 25% chance of having a normal child, could be explained by Harper's (1984) contention that parents frequently reverse the probabilities they are given. Thus a 1 in 4 chance of having an affected child is interpreted as a 1 in 4 chance of having a healthy child.

A substantial proportion of mothers (approximately 40%), fathers (approximately 30%) and patients and siblings (over 60%) had also failed to grasp the critical fact that, with autosomal recessive inheritance, all pregnancies carry a 1 in 4 risk of recurrence no matter how many affected children may already have been born.

Considering that the prenatal diagnosis for CF is in its early stages, it was not unexpected that over 20% of parents and patients and approximately 45% of siblings indicated that they did not know about amniocentesis. Respondents were similarly less well-informed regarding the accuracy of this test, with approximately 12% of parents erroneously believing that it was one hundred per cent reliable.

Correspondingly low levels of genetic knowledge have been reported by other CF researchers (McCrae, Cull, Burton and Dodge 1973; Allan, Townley and Phelan 1974 and Burton 1975). In their sample of CF families, Leonard and co-workers (1972) concluded that only half of the parents had the kind of comprehension which would make their genetic knowledge useful. Compared to the strong performance of parents and CF patients on disease and treatment questions, Nolan and his team (1986) also noted considerably less success on the genetics and reproduction items.

A limitation of the present study was its failure to assess respondents' knowledge of reproductive issues, particularly since Nolan et al (1986) found that 75% of male patients and over 50% of parents in his sample were unaware that most CF males are sterile. Given the finding that over 60% of parents in the present study had not been given information on their affected child's reproductive capacity, it is probable that levels of ignorance in this area would have been correspondingly high.

Whereas a lack of information may partly explain the poor performance of patients and siblings on genetics items (44.44% and 50% respectively had not been given information on how CF is inherited), similar figures for fathers (22.64%) and mothers (15%) alone cannot explain their low scores on many of the items. These latter figures do suggest that whatever information on genetics was provided, was either misunderstood or poorly retained. Another reason which might explain poor parental performance is ill-timed genetic counselling (McCrae, Cull, Burton and Dodge 1973; Falkman 1975 and De Wet 1984), particularly were it to have coincided with the emotional distress created by learning the diagnosis and prognosis. Because of the complex and intangible nature of genetic concepts, any information provided during this period is even less likely to be retained. Indeed, some researchers (Reynolds, Puck and Robinson 1974) found that at least 3 to 6 months were needed for couples to adjust to the discovery of a birth defect or genetic disease, before they were in a position to assimilate genetic information.

Evidence from a study of families with a child with congenital heart disease found that parental understanding and recall of genetic information may well be enhanced by the provision of a written description of recurrence risks (Halloran, Hsia and Rosenberg 1976). Written explanations can then be re-examined when and if the need arises.

5.2.4 Anatomy

Several researchers using both adult (Pearson and Dudley 1982) and paediatric (Kaufman and Hersher 1971 and Beales, Holt, Keen and Mellor 1983) samples have suggested that educating the patient about his illness and its treatment may be hampered by an inadequate knowledge of the anatomy and functioning of the body. In general, family members in the present study were well-informed about anatomy, although they correctly identified the stomach, pancreas and liver less frequently than the remaining body parts.

The successful performance of patients and siblings on these items may well be age-related. In their studies of healthy children, Gellert (1978) and Eiser and Patterson (1983) found that by approximately 12 years of age most children in their samples could identify the lungs, heart and stomach while knowledge of the liver was usually attained by early adolescence. Similarly, Harbord and co-workers (1987) reported that almost 80% of their older CF patients (12-18 years) knew that 2 organ systems were involved in the illness.

Furthermore, the finding in the present sample, that patients were more knowledgeable than siblings about both the location and function of the pancreas, is in keeping with those of Patterson and Tripp (1984a) who noted that significantly more diabetic patients than controls could correctly identify the pancreas. These authors speculated that diabetic patients may have placed more value on this organ than the control group by virtue of their illness experience.

Parents, on the other hand, although slightly less successful in correctly identifying the pancreas, in comparison to their knowledge of other body parts, were considerably more knowledgeable than a sample of adult surgical patients of whom less than 20% was able to locate the pancreas and explain its function (Pearson and Tait 1982). In contrast, most family members correctly identified the lungs.

Clearly an accurate knowledge of the location and function of the lungs and the pancreas is important if family members are to fully comprehend the nature of symptoms and the relevance of treatment. The relationship between body parts and the likelihood of compliance was explored in a group of children with juvenile chronic arthritis (Beales, Holt, Keen and Mellor 1983). These authors found that children who had a clear understanding of the internal manifestations of their disease were more likely to appreciate the purpose of therapy. They noted furthermore, that below 11 years their patients had immense difficulty in appreciating how tablets which were swallowed and hence entered the stomach could possibly affect painful joints in other parts of the body. Yet, in CF, the patient from an early age is required to understand the complicated relationship between a poorly functioning pancreas and the need for enzyme supplementation, the dosage of which is determined partly by the fat content of the food (i.e. what enters his mouth) and the presence of oil in the stool (i.e. what leaves his body). Moreover, children under the age of 9 years are unlikely to have yet linked the digestive and excretory systems (Gellert 1978 and Eiser and Patterson 1983).

Beales et al (1983) also found that the younger patients had great difficulty in recognizing that benefits could accrue from 'nasty' tasting medicines which had an immediate negative effect upon them. Notwithstanding the positive responses reported by Harbord and colleagues (1987), unless care is taken to explain, in detail, how and why physiotherapy clears the clogged bronchi of the lungs, the young CF patient might also fail to appreciate the benefits of this often poorly accepted form of treatment (Burton 1975 and De Wet 1984) .

As was suggested for genetics, written information in the form of simple age-appropriate drawings, may be a valuable adjunct to enhancing family members' basic understanding of anatomy and bodily function relevant to CF. Well-designed written information could also serve to satisfy the need, expressed by

50% of siblings, over 30% of parents and 22% of patients, for a great deal more information on how the body works.

5.2.5 Terminology

Less than half the sample of parents and patients and less than one-third of siblings correctly identified the terms dyspnoea, haemoptysis and steatorrhoea all of which are commonly occurring symptoms of CF. Since the panel of CF doctors had expected all parents to know the meaning of dyspnoea and haemoptysis, they had greatly overestimated parental knowledge of terminology (61.28% of parents failed to answer these two items correctly). Using a sample of adult patients with a variety of chronic illnesses, Byrne and Edeani (1984) also found that health professionals had significantly overestimated patient understanding of medical terminology.

Poor knowledge of medical terminology must also be viewed in the light of findings on the Index of Communication Problems. Approximately 28% of patients, 20% of mothers and 15% of fathers felt doctors used too many big words when providing explanations about the illness. A further 24% of fathers, 17% of patients and 13% of mothers often forgot what doctors told them about CF. These findings, together with those of Burton (1975) and De Wet (1984) that the excessive use of medical jargon leads to dissatisfaction among CF families, suggest that the health care team should not use language which may cause families to misunderstand or forget what they have been told. In similar vein, such usage should be avoided in all lay publications and should the use of medical terminology be unavoidable, a glossary of medical terms should be included in this material.

The broad implications for information-giving of findings from this section are addressed again in relation to family members' information needs (Section 5.5) and their sources of information (Section 5.6).

In addition to assessing family members' factual medical knowledge of the disease, the study also assessed their subjective perceptions of their child's present and future health. These findings are discussed in the next section.

5.3 PERCEPTIONS OF THE CF PATIENT'S PRESENT AND FUTURE HEALTH

Despite advancements in the survival rates of CF patients, it is still not possible to predict the course of the illness or the duration of survival for any particular child (Koocher 1984). Uncertainty about the future is, therefore, a significant source of stress among affected families (McCollum and Gibson 1970; Burton 1975 and De Wet 1984). The present study used two single item measures to assess family members' perceptions (i.e. 'knowledge') of the patient's present and future health.

Markedly positive perceptions of the CF child's present and future health were expressed by most family members. These findings are in keeping with those of previous CF researchers who also noted a tendency on the part of parents (Frydman 1981) and patients (Strauss and Wellisch 1981) to overestimate prognosis whilst retaining a fairly accurate perception of present health. Although estimates of present health in this study were high, they could not necessarily be interpreted as unrealistic given that 52.72% of patients were considered to be in 'excellent' or 'good' health according to their Shwachman ratings, with a further 25.42% falling in the 'mild' category.

In contrast, when seen against the medical realities of the disease, the finding that as few as 20% of parents and patients and 10% of siblings predicted any decline in the patient's health, could be considered somewhat optimistic. A similarly strong orientation towards life and living, despite the fact that there might not be a future, was noted by Susman, Hersh, Nannis et al (1982) in their study of seriously ill adolescents with cancer and their mothers. In yet another study involving children with cancer, parents and patients also appraised the future somewhat more optimistically than did the treating doctors (Mulhern, Crisco and Camitta 1981).

Various possibilities exist to explain these high ratings of present and future health. Strauss and Wellisch (1981) took their high ratings as evidence that their CF patients were using minimization (i.e. not making too much of the disease) in order to cope with an uncertain future. They saw minimization as existing on a continuum between preoccupation at one extreme and denial at the other. Similarly, Comaroff and Maguire (1981) contend that the processes involved in reaching an understanding of a fatal prognosis cannot be reduced to stable states definable as 'realistic acceptance or irrational denial' (p.118). Instead they believe that parents oscillate between contradictory responses which might include both optimism and despair. Certainly the open-ended responses of a significant proportion of parents and siblings suggested an underlying anxiety concerning the terminal nature of the illness, even if these fears were not immediately obvious from their scaled responses. (This discrepancy is addressed later in this section).

Beisser (1979) suggests that what appears, from a medical perspective, to be the denial of the realities of an illness may well represent the patient's affirmation of his health. In other words, the patient chooses to focus his attention on living as opposed to illness. For Beisser 'the patient may have an illness but his or her life would be of little value if that were all that the patient had' (p.1029). A similar response was noted by Venters (1981). In her sample, CF parents consciously opted to focus on the best qualities of the situation, rather than emphasize the child's potential lack of health. The lack, furthermore, of any significant correlations between perceptions of future health and clinically relevant medical variables (Shwachman score, days in hospital, number of clinic attendances) would further support a health rather than an illness orientation.

Although, for methodological reasons, the relationship between optimism and perception of future health was not formally examined, the high optimism ratings found among family members provided convincing evidence of a strongly positive orientation towards the future. Attitudes which, in all likelihood, would influence their perceptions of the patient's prognosis.

A more obvious reason for high ratings of the patient's Future Health would be a lack of relevant information about the life-threatening aspects of the illness. Over 50% of fathers and 60% of mothers, patients and siblings reported that they had not been given information on what to expect if the illness gets worse, while 35% of fathers, approximately 45% of mothers and patients and 68% of siblings had not been given information on the possible complications of CF. Indeed, Strauss and Wellisch (1981) felt that a possible reason for their CF patients' highly favourable perceptions of their future health may have resulted from 'unintentionally incomplete communication' (p.146) on the part of the doctors whom, they suggest, may have withheld information so as to allow the patients to maintain hope. It cannot, however, be discounted that parents (and for that matter patients and siblings) had been told about the potentially fatal aspects of the illness, but given the painful nature of such information, had failed to fully grasp its meaning (Burton 1975, Venters 1981 and De Wet 1984).

A word of caution is necessary when interpreting the high ratings of future health noted in this study. Rather than reflect overly optimistic perceptions for the future, these ratings may simply represent family members' reactions to the methods of assessment. Family members were required to acknowledge on paper that the CF patient might never get better and could, moreover, become a great deal worse - a task which, in all likelihood, proved too painful for most respondents who, as a result, opted for the less traumatic response options. Faced with a similar dilemma, Falkman (1977) asked her sample of mothers to rate their CF child's future prospects on a scale which included one hundred gradations. Despite a vastly increased range of response options, virtually no mothers in her sample were able to rate their child below fifty.

Further evidence to indicate that respondents, in the present study, may have had difficulty 'condemning' the patient to a hopeless future came from their responses to open-ended questions concerning the 'worst aspects' of the illness. The more indirect nature of these assessments enabled approximately 40% of fathers, 30% of mothers and 35% of siblings to express very real fears created by their 'knowledge' that the disease is incurable and inevitably fatal. However, even with an indirect form of measurement, only one patient was able to express death-related fears. It is noteworthy that in response to a similar question, Bywater (1981) and Harbord, Cross, Botica and Martin (1987) also found that only a minority of their CF patients referred to the fact that the disease was incurable, while the remainder of the respondents focused their concerns on problems of living with the illness.

Clearly a single item rating of future health, as used in this study, was not a reliable and valid measure of family members' perceptions of the patient's future health.

Notwithstanding the discrepancies in family members' responses, this discussion has highlighted a degree of ambiguity in their perceptions of the CF patient's prognosis. Alternatively, the findings could be interpreted as evidence of coping through the use of avoidance (Cohen and Lazarus 1980). In other words, while family members do not 'deny' the facts or implications of the illness, wherever possible, they choose not to talk or think about them. The medical team is, in turn, faced with the dilemma of to what extent family members should be apprised of the true facts (such as survival rates) about the illness. While in the past doctors argued that to confront parents and patients with the truth about a fatal illness would serve only to remove all hope (Wiener 1970), the most recent policy statement from the American Board of Pediatrics (1987) has advocated that parents and patients be given the whole truth about an illness. To this end doctors need to be equipped with the skills necessary to provide sensitive information, an issue which is addressed in Section 5.6.

5.4 INFORMATION PREFERENCES

The present study assessed how much family members wanted to be told about the condition so that information-giving would accord with the expectations of CF family members. A 3 item Index of Information Preferences, with acceptable psychometric properties, was used for this purpose. The mean Index scores for all family groups fell well above the mid-point of the possible range. The strongly positive responses in favour of complete openness toward information-giving provided convincing evidence that family members wanted and, indeed, considered it their right, to be kept fully informed about CF. These preferences were, furthermore, independent of any sociodemographic or medical criteria such as the patient's age or the severity of his condition.

These findings are similar to those reported by Cassileth and his colleagues (1980). These researchers interpreted the preferences of their adult cancer patients for open communication as evidence of a move away from the concept of the patient as a passive recipient of medical care, toward the more contemporary standard of a well-informed patient who actively participates in his own management. Comparative research among CF samples is not available. Nolan, Desmond, Herlich and Hardy (1986), however, anecdotally noted the incisive response of a 16 year old CF patient to the question 'what should patients know about CF?'. His reply quite simply began as follows: "They should know EVERYTHING' (p.235).

While the findings in this study can leave no doubt as to the wishes of most family members to be kept well-informed, there was, nevertheless, a minority who preferred not to be exposed to all the facts about CF. This suggests the need for an individualized approach in which the information preferences of family members are established prior to information-giving.

Such an approach would also support the contention that the right to know be counterbalanced by the right not to know (Madorsky, Radford and Neumann 1984). Information-giving should, furthermore, take cognizance of the specific information needs of individuals, the content of which was shown to vary greatly as a function of family grouping.

A final note of caution is necessary. While 85% of CF parents believed that they were entitled to all the facts about their child's condition, this study did not establish whether parents would necessarily concede that this right should be extended to their offspring. Indeed, parents might well believe that their children should be protected, for as long as possible, from facts such as prognosis and male sterility. Future research is needed to clarify the attitudes of both parents and clinicians in this respect so that information-giving can meet with the wishes of all parties concerned.

5.5 INFORMATION NEEDS AND INFORMATION NOT GIVEN BEFORE

Whereas there is ample evidence that CF families receive too little information about the illness (Burton 1975; Falkman 1977 and De Wet and Cywes 1985b), no studies have as yet addressed the content issues of the information being sought. Thus an important aim of the present study was to identify what family members wanted to know about selected medical and psychosocial issues. Family members also had to indicate whether this information had been provided in the past.

Based on their Index ratings, both fathers and mothers evidenced moderately high information needs. The mean ratings of fathers were, however, slightly higher than mothers. Moreover, when compared to mothers they wanted a significantly great deal more information on 35% of the specific items, the content of which was predominantly medical in nature. These findings were in contrast to those of Burton (1975) who found that fathers were less interested in acquiring further information particularly if they came from the lower social classes (III, IV and V). This discrepancy may reflect the nature of the two measuring instruments. Whereas Burton (1975) had assessed parental desire for further information by means of a single item, the present study used a more comprehensive instrument which reflected both total and specific needs and as such may have been a more sensitive and reliable measure.

Fathers were also consistently less likely than mothers to have been given medical information. Since fathers are less likely to attend clinic regularly and will therefore have less direct contact with the health care team, these findings were not unexpected. This interpretation received further support from the significant negative correlation (r_s - 0.38, $p = 0.0001$) found only between fathers' Index ratings of Frequency of Use of Informational Support and Information Not Given Before.

According to their Index ratings, patients evidenced the lowest Information Needs of all family groupings, in spite of the fact that more than 40% of patients had not been given information on at least three-quarters of the specific topics. These findings support those of previous research. In two studies (Boyle, di Sant'Agnese, Sack et al 1976 and Bywater 1981) researchers encountered strong resistance on the part of CF patients to knowing more about their illness, while Nolan and co-workers (1986) found that only one-third of their sample of patients wished to know more about CF.

The composite Information Needs of siblings were slightly higher than those of the patients. Siblings, too, had not been given information on many medical and psychosocial topics. Indeed, at least half the sample had not been given information on almost 85% of the items. Comparative data from other CF studies on siblings' information needs are not available.

Findings in the present study clearly indicated that whilst family members, and more particularly parents and patients, had been given medical information about CF, a large proportion remained uninformed regarding the psychosocial implications of the illness. These findings confirmed those of previous CF research (Frydman 1981). It was, therefore, not surprising that most family members wanted a great deal more information about the non-medical aspects of the illness.

New information about CF from other parts of the world, was the item most frequently endorsed as requiring a great deal more information by fathers, mothers and patients (approximately 80%, 70% and 60% respectively). Half the sample of siblings also wanted much more information in this area. Yet less than 50% of parents and 20% of patients and siblings had actually been provided with this information. Pfefferbaum and Levenson's (1982) finding that doctors underestimated adolescent cancer patients' desire for information about cancer research, suggests that health care professionals may not realize the extent of family members' wishes for this information, the more so given the technical nature of much of this material. No doubt the need to be kept informed of current developments in CF research was partly motivated by the hope that a cure for CF might soon be found and that more effective forms of treatment might have been discovered. Doctors need to recognize and address this strongly felt need.

Marked differences were evident between parents and patients and siblings regarding the content of the psychosocial information which they desired. Whereas only a small proportion of patients and siblings wanted a great deal more information about how to cope with feelings about the illness and how to discuss the illness with other people (e.g. parents, peers), these topics were endorsed by a substantial proportion of parents as requiring a great deal more information. Similarly only a minority of patients was interested in knowing how other CF patients cope with their illness. In contrast, most family members shared the need for a great deal more information about the future. This included a desire for 'positive' information about the patient's career, marriage and social life as well as 'pessimistic' information about what to expect if the illness deteriorates.

These findings share much in common with those of Levenson and her co-workers (1983) who noted a similar tendency, among a sample of adolescent cancer patients, to avoid information with personal and emotional connotations whilst wanting information about the future. These authors suggested that their patients might have been using avoidance or minimization to cope with their illness. Given the additional data in the present study that less than 30% of patients wanted a great deal more information about the complications of CF and that only one-third of patients wanted much more information about how serious their illness was, they could also have been using similar coping mechanisms. (It has already been suggested in Section 5.3 that the very positive attitudes of patients towards their future health, could well reflect a tendency to deny illness and affirm health).

Given the accumulated evidence (Turk 1964; Meyerowitz and Kaplan 1967; McCollum and Gibson 1970; Burton 1975 and De Wet 1984) attesting to the difficulties which CF parents experience when discussing disease-related issues, it was not surprising that fathers and mothers required much more information on how to discuss the illness with their children, particularly where such discussions involved the possibility of the patient's death.

Information regarding the patient's reproductive capacity was highly sought after by almost two-thirds of parents, while over 40% of siblings wanted a great deal more information regarding their chances of producing affected offspring. A similar desire to know more about carrier status was noted among siblings in a small sample of CF families in which a child had died from the illness (Kerner, Harvey and Lewiston 1979). The need for further information on carrier status was also found amongst siblings of children with Duchenne muscular dystrophy (Firth 1983 and Fitzpatrick and Barry 1986).

Several possibilities exist to explain why a large proportion of family members had not been given psychosocial information. Given the finding that approximately 11% of the variance in parental Index ratings of Information Not Given Before was determined by their Index ratings of Doctor-Parent Communication Problems, it may be assumed that some parents could have experienced difficulty in raising such issues with the clinic doctors. Certainly the perceived lack of privacy at the clinic, experienced by a significant minority of parents, could well have deterred them from initiating discussion of sensitive topics. Although the item did not refer specifically to psychosocial information, almost 25% of fathers, 17% of patients and 13% of mothers indicated that they often forgot what the doctors told them about CF. Thus it is plausible to speculate that family members may have been given psychosocial information which, because of its potential for raising anxiety, they had failed to retain (Burton 1975).

Given the two-way nature of the communication process (Pritchard 1986), it is possible that factors within the doctors (e.g. attitudes and skills) may have partly determined what and how much psychosocial information was given to families. For example, they may have felt ill-equipped to counsel families on sensitive, emotionally-charged issues (Wolraich and Reiter 1979 and Sack, Fritz, Krener and Sprunger 1984). Doctors might also have believed that family members, and more especially patients, should be protected from knowing all the details about CF (Wiener 1970). Furthermore, it is possible that the doctors based their information-giving on what they believed families wanted to know (Waitzkin 1984), perceptions which are not always in accord with those of the recipients of the information (Pfefferbaum and Levenson 1982 and Strull, Lo and Charles 1984). Since the panel of clinic doctors had overestimated parents' medical knowledge of CF, they may also have overestimated their knowledge of psychosocial issues and thus provided less information.

Given the constant time constraints that operate during clinic visits, the immediacy of the patient's clinical status might have limited the exchange of information to medical issues. The multidisciplinary nature of the CF clinic could also have led team members to mistakenly believe that psychosocial information had already been shared with parents and patients. Several studies (Taylor and Merrill 1970 and Pless, Satterwhite and Van Vechten 1978) have highlighted the lack of clear responsibility among team members as to who should provide what information to patients and their families.

That a substantial proportion of mothers, themselves a main source of information for fathers, patients and siblings had not been given information on the psychosocial implications of the illness, could partly explain why correspondingly high numbers of family members had not received this information. It is also possible that patients were inhibited from pursuing sensitive issues because they perceived a reluctance on the part of parents to engage in such discussions (Waechter 1971).

A discussion of the implications for information-giving of the findings in this section follows. Clearly, CF families should be receiving a great deal more information on medical and, more especially, psychosocial issues. To the extent that a knowledge of physiotherapy, nebulization, pancreatic supplementation and nutrition is a prerequisite for optimal adherence to the treatment regimen, it can be considered essential knowledge. As such, this information must be given to all parents and patients regardless of expressed need. For their part, siblings would benefit from more medical information about the treatment of CF which should include guidelines on suitable forms of exercise and sport for CF patients. Commonly held misconceptions in family members' understanding of CF need to be identified and removed.

Nor can the health professionals assume that because they have supplied this information before that it has been understood or retained. On the contrary, if one is to assume that the clinic doctors had based their projected assessments of parental performance on the Revised CF Knowledge Test (Section 3.3.1) on their conviction that all the information on this Test had actually been provided to parents, it would seem that parents failed to retain approximately 24% of the material (30% of which included such crucial information as respiratory symptomatology and treatment, nutrition and genetics).

Alternatively, the clinic doctors may have overestimated their information-giving behaviour (Waitzkin 1984). This latter explanation is particularly worrisome as doctors are unlikely to take the initiative in providing information which they firmly believe parents already possess. It would seem, therefore, that doctors should heed the advice of Pless and his co-workers (1978): 'As a rule parents must be asked explicitly and regularly if they understand explanations or instructions and should routinely be asked if they have any questions' (p.16). Since increasing numbers of CF patients are reaching early adulthood (Mearns 1986) and will be assuming primary responsibility for their medical care, the health care team will also need to be particularly alert to gaps in the medical knowledge of this group of patients.

That CF families should also be receiving anticipatory guidance on medical and psychosocial issues was underscored by their expressed need for a great deal more information about career opportunities, marriage, reproduction, complications and what to expect if the illness gets worse. While it has been argued that such information serves only to increase anxiety and despair (Jacobs 1977), several CF authors are adamant that to withhold such information is to give unrealistic and false hopes about the future (Sibinga, Friedman and Huang 1973 and Strauss and Wellisch 1982). Power and Sax (1982) found, furthermore, that having a knowledge of what to expect prepared neurological patients and their families to cope with periodic medical problems and led to a reduction in the number of unscheduled visits to the clinic. As one parent in the present study commented in relation to an episode of meconium ileus equivalent '... because we don't know enough it is a natural reaction to panic... whatever I know is due to experience and not knowledge. And it would be a lot easier if we had both knowledge and experience.'

Whereas most parents of healthy children can rely on their childhood experiences of being parented as well as on the advice of friends, relations and neighbours, these sources are generally inadequate for parents of children with special needs (Wikler 1981). Thus, in the same way as anticipatory guidance is a routine part of well child supervision in general paediatrics so, too, should families with a chronically ill child be given information about critical periods (e.g. school entry, hospitalization) or issues (e.g. infertility, delayed menstruation) in advance so that they can plan for areas of expected difficulty (Mearns 1986 and Walker, Ford and Donald 1987).

The tendency for many CF patients in this study to avoid certain information, again underscores the need for a policy of information-giving adjusted to the preferences and coping patterns of each patient. A similar tendency towards minimization in adolescent cancer patients, prompted Levenson and her colleagues (1983) to recommend that patients' information needs be reassessed at regular intervals to determine current levels of disease-related knowledge and their readiness to receive additional information. Such an approach to information-giving would both accord with the cognitive-developmental needs of these patients (Burbach and Peterson 1986) and would increase the relevance of the information provided.

The main sources of family members' medical and psychosocial information are discussed in the following section.

5.6 SOURCES OF INFORMATIONAL SUPPORT AND GUIDANCE

The task of accumulating knowledge about CF can be either facilitated or impeded by the availability and quality of informational support (Chesler and Yoak 1984). One of the aims of the present study was to identify and evaluate important sources of information and guidance used by CF family members. In this section findings relating to the following measures of informational support are discussed: ratings of Frequency of Use and Perceived Helpfulness and Doctor-Parent/Patient Communication Problems. The relationship between family members' CF knowledge and their sources of informational support is also addressed.

In the course of coping with a lifelong illness like CF, family members are likely to consult a number of professional and lay sources of information. According to mean item ratings, one's spouse or parents, the clinic doctors, prayer and faith, and books, pamphlets and magazines about CF were the most frequently used sources of informational support and guidance. In keeping with the correlational evidence provided by the two Index ratings, these sources were also rated by parents and patients as being the most helpful.

These findings are consistent with previous research which noted that family members relied most upon doctors (Allan, Townley and Phelan 1974; Burton 1975; Mikkelsen, Waechter and Crittenden 1978; Bywater 1981; Frydman 1981; Strauss and Wellisch 1981 and Phillips, Bohannon, Gayton and Friedman 1985) and mothers (Strauss and Wellisch 1981 and Nolan, Desmond, Herlich and Hardy 1986) for their information about CF.

However, in contrast to previous CF findings (Blumenthal 1969; Burton 1975 and De Wet and Cywes 1985b), parents and patients in the present study perceived their encounters with the doctors as being relatively free of communication problems. Most respondents were highly satisfied with the clarity of information and the honest manner in which it had been provided. The use of medical jargon, the lack of privacy at the clinics and the tendency to forget information were, nonetheless, problematic for a minority. The implications of these latter problems have already been discussed in Sections 5.2 and 5.5.

Caution is needed when interpreting these high levels of satisfaction with the communication process. Although care was taken, when constructing the Index, to reduce bias caused by acquiescent response set (Ware 1978) and wanting to please the interviewer (Lebow 1974), parents and patients may still have been reluctant to criticise the doctors upon whom they depended for their medical care (Locker and Dunt 1978). The Index may also have failed to include items which were better indicators of perceived communication problems (Gutek 1978). Finally it is possible that the low Index ratings were saying more about the respondents' expectations than about the actual services they were receiving. Locker and Dunt (1978) suggest that although a service (in this case, the exchange of information) does not fully meet their needs, the recipients rationalize that the providers (i.e. the doctors) are doing all that can be expected under the circumstances. Thus although parents and patients still had many unmet information needs, they may have believed that doctors were doing their best (Woolley, Kane, Hughes and Wright 1978).

As frontline providers of information to CF families, doctors will need to be equipped with the skills necessary to impart both medical and psychosocial information. Given, furthermore, the painful nature of much of what CF families want and need to know, doctors will require skills which enable them to focus on both the what (i.e. content) and how (i.e. manner) of information-giving (Wolraich and Reiter 1979). Such skills should, in turn, alert them to cues as to the readiness of individual family members to learn more about the illness. Since the Information Preferences of a minority of family members indicated a certain reluctance to being given all the facts about CF, the need for an individualized, empathic approach becomes even more imperative. The importance of a sympathetic and hopeful attitude on the part of the doctor when imparting information has also been stressed by previous CF research (Burton 1975 and De Wet and Cywes 1985b).

While most research on coping has, to date, focused recipients of informational support, far less attention has been paid to the providers of such support (Wortman 1984). Anecdotal (Axelrod 1978) as well as empirical (Lewiston, Cowley and Blessing-Moore 1981) findings suggest that CF doctors experience considerable stress in the course of their duties. Thus any failure to equip them with the skills necessary to fulfil their role may simply add to an already stressful situation.

The finding that fathers, patients and siblings all selected mothers as their main source of information about CF, also has important practical implications for the doctors. Since mothers depend mainly on clinic doctors for their information they, in turn, will need to ensure that mothers possess both the knowledge and the skills to impart this information to their families (Nolan, Desmond, Herlich and Hardy 1986). Although mothers may cope adequately in providing information of a less sensitive nature, discussions incorporating such topics as prognosis and sterility may prove too threatening (Turk 1964; McCollum and Gibson 1970 and De Wet 1984). Recognizing that this could be a potential problem for some CF parents, Tropauer and his team (1970) suggested that doctors may need to involve themselves more directly in educating adolescent and young adult CF patients about their future.

Of practical interest was the counter-intuitive finding that most CF patients preferred to have their parents present during consultations with the clinic doctors. Although this finding needs replication among a far larger sample of patients of varying ages it did, nevertheless, confirm similar sentiments expressed by a sample of adolescent cancer patients (Levenson, Pfefferbaum, Copeland and Silberberg 1982).

No doubt their less frequent contact with hospital-based sources made fathers more reliant on mothers for their CF information. The finding that patients and siblings depended more on their mothers than fathers for CF information may have resulted from traditional role divisions within the family. Fathers may simply have been at home less often than mothers and were therefore less available for disease-related discussions. Since fathers were only marginally less knowledgeable than mothers about the medical aspects of CF, they should be encouraged to play a more active role in educating patients and siblings about the disease. This could also have broader implications as Venters (1981) found that where CF families were able to share the burden of coping with the illness, levels of family functioning were significantly higher than among families in which a single member assumed the total burden of care.

Prayer and faith were also highly rated (2nd and 3rd rankings) by fathers, mothers and patients as being both frequently relied upon and as providing quite a lot of helpful information and guidance about CF. The importance of personal faith as a source of guidance in coping with CF has been noted by several CF researchers (Mikkelsen, Waechter and Crittenden 1978; Venters 1981 and De Wet 1984). Given the life-threatening nature of the illness which inevitably gives rise to many existential questions, these findings were not entirely unexpected. The more so since Western medicine tends to be more intent on solving questions related to the 'how' rather than the 'why' of illness (Comaroff and Maguire 1981). Thus while clinic doctors are particularly well suited to providing bio-medical explanations for a disease, they remain ill-equipped to answer questions concerning ultimate causation (Hiatt 1986), such as why a particular family has been singled out to suffer irreversible tragedy.

Faced with events for which no rational explanation seems forthcoming, family members may exhibit what Chodoff, Friedman and Hamburg (1964) have described as 'an existential hunger for a meaningful and understandable explanation' (p.747) of a seemingly inexplicable situation. Faith and prayer may well enable a sizable proportion of CF parents and patients to make spiritual sense of their ostensibly meaningless predicament. Moreover, from a coping perspective, personal faith fulfils a dual function. On the one hand it influences how the stressor (i.e. the life-threatening illness) is appraised and secondly, it serves as a source of hope which facilitates overall coping with the illness (Barbarin 1983).

Irrespective of the exact nature of the guidance obtained through personal faith and prayer, it is a highly valued source of support which fulfils an important function complementary to that of clinical medicine. While the doctors meet the factual information needs of family members by enhancing their medical knowledge of the disease, faith and prayer facilitate understanding of the meaning of life itself.

Given these findings, the correspondingly low utilization and helpfulness ratings for the priest or minister were somewhat surprising, although a similar discrepancy has been recorded among a sample of parents with a child with cancer (Morrow, Hoagland and Morse, 1982).

Several parents felt that their priest was generally ignorant concerning the facts of the disease and was, instead, dependent on them for CF information. Similar sentiments were expressed by a mother in Burton's (1975) sample who commented 'They're (ministers) very sympathetic but they don't understand' (p.60). Limited contact with the priest or minister may be a further factor inhibiting family members' use of this source of information and guidance (Morrow, Hoagland and Morse 1982). Personal faith, on the other hand, because it constitutes an internalized belief system, is a readily available source of guidance. Similarly, prayer provides a direct link with God or some other supreme being. However, these interpretations by their very nature remain speculative.

Books, pamphlets and magazines about CF were also consulted quite often and found to provide some helpful information and guidance. Previous research concerning the value of written information about CF has produced conflicting viewpoints. Parents in Burton's study (1975) were generally satisfied with the CF literature, so much so that fathers rated written material as being more informative than doctors. Nolan and his team (1986) also found that a substantial minority of parents relied upon 'medical books' for their CF information. In contrast, other CF studies (Lawler, Nakielny and Wright 1966; Allan, Townley and Phelan 1974; Strauss and Wellisch 1981 and De Wet and Cywes 1985b) noted considerable dissatisfaction with the published CF literature. Respondents in these studies felt that too much emphasis had been placed on the negative aspects of the disease. No doubt the variable quality of much of the published literature was partly responsible for these discrepant findings. Certainly this seemed to be so in the present study. Whereas parents were full of praise for the 'overseas' literature, they were highly critical of the local publication which they felt was too technical and pessimistic.

While the written word cannot be seen as a substitute for the verbal sharing of information, there is no doubt that well-prepared literature can serve as a valuable adjunct to face-to-face communication (Van Vechten, Satterwhite and Pless 1977). The important role of written material was further underscored by the finding that fathers who had consulted the CF literature 'many times' had significantly more CF Knowledge than fathers who had used it only 'occasionally' or 'not at all'. Since fathers were also less likely to have direct contact with hospital-based sources of information, the potential contribution of written CF material to their overall understanding of the illness, is underscored still further.

CF literature can, furthermore, be particularly beneficial during times of crisis, such as the diagnostic period, when CF parents often fail to retain what they are told (Burton 1975). Moreover, evidence that parents are least likely to remember the clinical details concerning a chronic condition (Kupst, Blatterbauer, Westman et al 1977) suggests that written information, incorporating technical details (e.g. inheritance patterns) may be an indispensable supplement to the verbal exchange of this information. Finally written material can serve as a valuable medium for keeping families abreast of developments in other CF centres worldwide.

Previous research concerning the value of the CF Association as a source of information and guidance, has also yielded conflicting results. In keeping with the moderately useful assessments made by parents in the studies of Burton (1975) and Phillips, Bohannon, Gayton and Friedman (1985), parents (and more particularly mothers) in the present study also rated the CF Association as offering a small measure of helpful information. Patients, on the other hand, neither relied much on the Association for their information, nor had they found it particularly helpful. In contrast, over 70% of mothers in Bywater's (1981) sample said that they had learnt more about the illness from the CF Research Trust than from the medical team.

Yet other findings from an Australian sample (Allan, Townley and Phelan 1974) indicated that two-thirds of the CF mothers were adamant that they would not join the Association because they were perfectly satisfied with the information they were receiving from the hospital. Again these reported differences between samples may well be attributable to the variable quality and breadth of informational support offered by these organizations.

Certainly the findings in the present study, that fathers and mothers who had no contact with the CF Association had significantly less CF Knowledge than parents who enjoyed occasional or very frequent contact, suggest that the Association has an important role to play in enhancing understanding of the illness. A word of caution is necessary when interpreting the relative value of the CF literature and the CF Association. Since a main function of the Association is, in fact, the dissemination of written information about CF, it becomes very difficult to evaluate the independent contribution of the CF Association to family members' knowledge of the disease.

While these findings have highlighted the importance of the CF literature and the CF Association as efficient channels for the transmission of factual medical information, unless they are made accessible to CF families from all social classes, their maximum benefit is unlikely to be felt by those groups who are most in need of further information.

Other sources of information to receive moderately low frequency and helpfulness ratings were other parents with a CF child and the family doctor. Only a minority of fathers and mothers (7.41% and 20% respectively) had obtained a great deal of helpful information about CF from other parents with a CF child, while no patients had received such help from fellow patients.

Although some parents (Driscoll and Lubin 1972 and McCollum and Gibson 1972) and CF patients (Strauss, Pedersen and Dudovitz 1979) have reportedly learnt a great deal about the illness and about coping in general through sharing experiences with each other, yet other samples of parents (De Wet 1984) and patients (Strauss and Wellisch 1982) have found these sources decidedly unhelpful. Based on research among families with a child with cancer, Comaroff and Maguire (1978) noted that other parents and patients were most helpful at points of crisis, such as the initial diagnosis and hospitalizations, whereas during times of relative stability, these people often acted as sources of 'disorienting' information. Comments to this effect were made by respondents in the present study who reported that their fears were often expanded rather than reduced by discussions with similarly affected families. Furthermore, where parents are themselves ill-informed, there is the ever present danger of exchanging false or inaccurate information (Rowland and Armstrong 1983).

These disparate findings suggest a high degree of individuality in the way CF families appraise this source of support. Thus any decision to pursue further contact among similarly affected families, is probably best left to the discretion of the individual parties. Notwithstanding the drawbacks of self-help, the mutual exchange of experiences has the potential for being a valuable source of both informational and emotional support for some CF families.

Less than 15% of mothers and 10% of fathers and patients had frequently approached their family doctor for information about CF, while less than 4% of family members had obtained a great deal of helpful information from this source. The limited supportive role of the general practitioner (GP) has been noted in previous CF research (Burton 1975 and Frydman 1981) as well as in studies conducted among other chronic illness groups (Pless, Satterwhite and Van Vechten 1978 and Stein, Jessop and Reissman 1983).

In general CF parents, together with a few patients, did not feel that the GP's possessed enough specialist knowledge to enable them to counsel a family about CF. Moreover, where a GP had failed to make the initial diagnosis or had been responsible for a series of misdiagnoses, parents' comments were even more scathing and they were even less able to view the GP in a positive light. The dramatic improvement which inevitably followed a correct diagnosis and the institution of suitable treatment, no doubt provided parents with still further evidence of the 'superior' knowledge of the clinic doctors.

It is also possible that given the severity and relative rarity of CF, together with inadequate educational preparation for managing chronic illness (Harding, Heller and Kessler 1979), GP's may have willingly opted to relinquish responsibility for these children in favour of hospital-based specialist care. While it cannot be denied that most GP's do not possess technical knowledge or skills on a level equivalent to that of the CF sub-specialist, the family doctor is nevertheless in a position to offer informational support of a more general nature, particularly regarding psychosocial issues and future planning, areas which are notoriously neglected in tertiary care (Frydman 1981). This would, however, entail close cooperation between primary and tertiary care practitioners, along with a regular exchange of medical and laboratory information concerning the CF patient's current medical progress. Furthermore, such a shared approach 'requires both the generalist and the sub-specialist to have a mutual recognition of and respect for each other's expertise, as well as agreement on the total care needs of the child' (Stein and Jessop 1984, p.190). To this end it is hoped that the recent introduction of a programme aimed at teaching primary care paediatric residents the prerequisites and skills required to manage chronic paediatric illness (Desguine 1986) may prompt other teaching institutions to include similar courses in their curricula.

Since older siblings are likely to have a minimum amount of contact with clinic doctors, one could speculate that the family doctor might have a valuable role to play in meeting their information needs.

Although hospital physiotherapists were the most frequently used paramedical members of the multidisciplinary team, their mean ratings of frequency of use and perceived helpfulness were nevertheless low. Considering that only a minority of fathers (26.42%), mothers (16.67%) and patients (5.56%) felt they needed a great deal more information about physiotherapy, it was not surprising that they had not had much recent contact with this source. However, the finding that a high proportion of parents and patients incorrectly believed that antibiotics are best given via a nebulizer before physiotherapy, suggests that their knowledge on treatment-related issues cannot be taken for granted. Since parents and patients may not be aware of the gaps in their knowledge, the onus rests with the physiotherapists to conduct periodic evaluations of their understanding of basic physiotherapy principles. Physiotherapists are also ideally suited to provide information on suitable forms of exercise and sport for CF patients - information which was highly sought after by approximately one-half and one-third of fathers and mothers respectively.

Parental ratings indicated that the dietician, the local pharmacy and the clinic social worker were the least used and least helpful sources of information and guidance about CF. Several explanations are possible to account for these findings.

Because most families obtained their medicines directly from the hospital, few had felt the need to consult their local pharmacist. Written comments further suggested that parents did not think that the pharmacist knew much about CF anyway.

Since the CF clinic did not encourage special diets as part of the treatment regimen, parents may not have been informed as to the availability, on the team, of a dietician. And considering that the clinic doctors were unaware that so many parents erroneously believed that fats should be excluded from the diet, they probably did not feel that nutritional counselling by a dietician was necessary. Thus when questions arose concerning dietary matters, in all likelihood, these were answered by the doctors themselves.

Considering that approximately 50% of fathers and 35% of mothers wanted a great deal more information about healthy meals for CF children, with a further 45% of patients needing much more information on what foods to eat to increase their weight, it would seem that the dietician does have an important educative role to play in the management of the CF patient. Against the generally poor performance of family members on questions requiring the application of nutritional knowledge, together with the well-described problems of undernutrition and low energy intake among CF patients (Dodge 1985), the potential value of the dietician is underscored still further. According to their written comments, parents and patients were ill-informed as to the role of the dietician. The onus would seem, therefore, to rest with the dietician to initiate contact with individual families. Clinic doctors could also act as an important referral source.

The low item ratings of use and perceived helpfulness of the hospital social worker are in keeping with previous CF research (Harrison 1977; Bywater 1981 and De Wet 1984), although these latter studies did not specifically focus on the social worker as a source of CF information. Findings in the present study, like those previously cited, suggest that parents and patients were under the mistaken impression that contact with a social worker is only justified in the presence of a 'legitimate ' social problem. Parents and patients appeared unaware that the social worker is well-equipped to offer psychosocial information and guidance. Since doctors are unlikely to raise psychosocial issues during clinic visits (Lau, Williams, Williams et al 1982), the contribution of the social worker assumes even greater importance.

The onus would seem, therefore, to lie with the social worker to enlighten families as to what informational support he/she can offer. By focusing on the educative component of her role, it might also serve to reduce the social stigma frequently attached to social work intervention.

A recent study noted that doctors are more likely to positively evaluate 'non-physician health professionals' (i.e. dieticians, social workers and physiotherapists) if their medical training has included exposure to the functions of the multidisciplinary team (Lasswell and Smith 1987). This finding suggests that medical educators would be well advised to include members of the allied health professions in their training programmes from an undergraduate level. Such exposure might, in turn, lead to greater cooperation between the medical and paramedical professions in the management of chronically ill patients.

When recommending a multidisciplinary approach to the distribution of CF information, a note of caution is necessary. Research, among parents with a child with congenital heart disease, has shown that parents prefer to obtain all their information directly from the medical specialists, through fear that non-medical sources may lack the knowledge to answer their questions (Kupst, Dresser, Schulman and Paul 1976).

A further problem, though not well documented in the CF literature, concerns the lack of coordination amongst the various sources of information in a multidisciplinary team; a problem compounded by the lack of clear-cut parameters concerning who is authorized to give what information to families (Mathews 1983 and Pritchard 1986). Pless, Satterwhite and Van Vechten (1978) recommended that one person, ideally the treating paediatrician, should become a definitive source of information. His task would be to interpret and coordinate information given to family members so as to avoid overlap and to ensure that family members actually receive the information they require. Regrettably, few busy specialist clinics meet this ideal. A decidedly more practical solution might be to include a checklist in each CF patient's folder, which could reflect what, when and by whom information has been given to a family (Dunkelman 1979).

5.7 SOCIODEMOGRAPHIC AND MEDICAL CORRELATES OF CF KNOWLEDGE, PERCEPTION OF PATIENT'S PRESENT AND FUTURE HEALTH AND RATINGS ON THE INFORMATION MEASURES

Empirical research from a wide spectrum of chronic paediatric disease has produced inconclusive evidence concerning the relationships between disease-related knowledge and sociodemographic and medical variables (see Literature Review: Sections 3.1, 3.2 and 3.1.2.2). One of the aims of this study was to examine the correlates of CF Knowledge and the various measures of Information, notably Information Preferences, Information Needs, Information Not Given Before and Sources of Information.

5.7.1 Age and Education

The age of the patient was not significantly associated with parents' CF Knowledge, Perception of the patient's Present and Future Health or their performances on the Index measures of Information. Age was, however, significantly associated with one specific item of information. Parents with an older CF child were significantly more likely to have enough information on How to use a nebulizer and to have been given this information before. Since persistent pseudomonas colonization is more likely in older CF patients for whom nebulized antibiotics may become the treatment of choice, these findings were not unexpected.

In similar vein, patients' and siblings' Composite CF Knowledge was not significantly associated with their age. These findings were in contrast to those of Nolan, Desmond, Herlich and Hardy

(1986) who reported that age was a significant predictor of their CF patients' knowledge. Different measures of CF knowledge could have accounted for these discrepancies. Older patients (14-22 years) did, however, obtain consistently higher average scores on 8 out of 9 sub-sections. On 2 of the sub-sections, viz. General CF Knowledge and Nutrition, these differences were significant. These trends supported the accumulated evidence that older (Hamburg and Inoff 1982; Johnson, Pollak, Silverstein et al 1982 and Susman, Hersh, Nannis et al 1983) and more cognitively mature (Beales, Holt, Keen and Mellor 1983 and Allen, Afflek, Tennan et al 1984) chronically ill children are likely to have a better understanding of their illness and its treatment.

The significantly superior knowledge of older CF patients on nutritional items may also have resulted from more trial-and-error experience with different kinds of food. However, since children from as early as the pre-teens are likely to begin taking greater control over their diets, eating more snacks between meals (Singleton and Rhoads 1982) and will be drawn towards less nutritional foods (Lamme and Lamme 1980), greater emphasis needs to be placed on equipping younger patients with the knowledge necessary to ensure optimal nutritional management.

Findings concerning the relationship between siblings' age and their CF Knowledge were counter-intuitive. Younger siblings exhibited higher levels of CF Knowledge on the Composite Test and on 6 out of the 9 sub-sections. Reasons for these findings are unclear. Since female siblings obtained higher scores than male siblings it was possible that females may also have been younger than their male counterparts. This proved not to be the case. It is possible that younger siblings spent more time at home and may therefore have experienced more direct contact with the patient and his management. Older siblings, on the other hand, might have been preoccupied with activities outside the home. Without comparative data these interpretations remain highly speculative.

Although low and scarcely significant, the correlations between patients' Index ratings of Doctor-Patient Communication Problems and their age ($r_s = -0.44$, $p = 0.01$) and education ($r_s = -0.47$, $p = 0.02$), warrant consideration because of their implications for information-giving. Several authors (Perrin and Perrin 1983 and Carraccio, McCormick and Weller 1987) have stressed that optimal communication of medical information between patients and doctors depends on the degree of 'match' between the information provided and the sick child's level of understanding. Information that is too simple may be dismissed (Perrin and Perrin 1983), whereas information that is beyond the child's level of understanding is likely to be misinterpreted (Brewster 1982) or may heighten the child's anxiety (Blos 1978).

Carraccio and her co-workers (1987) also warn that "...the pediatrician's appraisal should not be altered by an appearance of superficial sophistication that some children may acquire as part of their experience with chronic disease" (p.982). It is important, therefore, that medical staff use language (Eiser,

Eiser and Hunt 1986) and explanations (Whitt, Dykstra and Taylor 1979; Pantell and Lewis 1983 and Potter and Potter 1984) which are appropriate to the sick child's level of conceptualization. Almost 30% of patients in the present study felt that the doctors used too many big words. Since the sick child's conceptualization of his illness, treatment and future is likely to change in line with his advancing cognitive abilities (Beales, Holt, Keen and Mellor 1983 and Carraccio, McCormick and Weller 1987), information-giving must also keep pace with his improved level of understanding.

Paediatricians, however, may not always be familiar with children's cognitive development. Perrin and Perrin (1983) found that paediatricians tended to approach all children as if they were functioning according to the Piagetian stage of concrete operations (i.e. middle childhood). Thus they tended to overestimate the younger child's conceptual understanding of his illness, while they underestimated the older child's comprehension. These findings suggest that doctors need to pay more attention to developmental issues when providing information to their chronically ill patients if "...frustration and miscommunication" (Perrin and Perrin 1983, p. 878) are to be avoided and the information retained and fully comprehended.

5.7.2 Gender

An association between gender and levels of disease-related knowledge has been noted in previous CF research. Findings have highlighted the superior knowledge of mothers (Burton 1975 and Nolan, Desmond, Herlich and Hardy 1986) and female patients

(Nolan, Desmond, Herlich and Hardy 1986). The accumulated evidence was further substantiated in the present study. CF mothers and female siblings obtained significantly higher Composite CF Knowledge Test scores than fathers and male siblings respectively. Female patients also obtained higher Composite scores than male patients but the difference between their performances fell slightly beyond the accepted significance level (p=0.06). Female patients and siblings also exhibited superior knowledge on 8 out of 9 sub-sections on the Test.

Despite the consistently better performances of mothers and female patients, interpretation of these findings in the literature remains speculative. Burton (1975) suggests that because of role expectations in Western society, mothers assume responsibility for the medical management of the CF patient and this requires them to have adequate knowledge of the illness. As was the case in this study, mothers are also more likely than fathers to rely on the clinic doctors for their information about the illness (Burton 1975).

The superior knowledge of female diabetic patients has been tentatively ascribed to the greater developmental maturity of girls and to the likelihood that they spend more time with their mothers who are themselves better informed about the illness (Johnson, Pollak, Silverstein et al 1982). Traditional sex role expectations were probably also implicated in the significantly superior nutritional knowledge of female patients and siblings.

Gender was not significantly associated with parents', patients' or siblings' Information Preferences. Differences between the global and individual item ratings of fathers and mothers on the Index measures of their Information Needs, Information Not Given Before and Sources of CF Information have already been discussed in Sections 5.5 and 5.6. No significant gender differences were found for patients and siblings on these measures. The differential effect of social class on fathers' and mothers' Index ratings of their Information Needs, Information Not Given Before and Doctor-Parent Communication Problems is also addressed elsewhere (Section 5.7.4.).

5.7.3 Marital Status

Only 6 mothers fell into the category of single parents in this study. Thus while they obtained significantly lower scores than married mothers on the Composite CF Knowledge Test, reliable interpretation of this finding was not possible. Marital status, as an independent variable, was therefore dropped from further analyses in the study. Until the effect of marital status, on both CF knowledge and information needs, is investigated in far larger samples of single parents, it is only possible to caution health care professionals that, as a group, single parents may know less about the illness than their married counterparts.

5.7.4 Language, Race and Social Class

Social class proved to be the most significant explanatory variable in this study. Findings showed that parents from the higher social classes (I and II) had consistently increased CF knowledge, lower information needs, had been given more information about CF and were less likely to experience problems in communicating with clinic doctors than parents from social classes III, IV and V. In statistical terms, social class had respectively been responsible for 53.47%, 15.84%, 22.38% and 12.93% of the variance in parental CF Knowledge and their Index ratings of Information Needs, Information Not Given Before and Doctor-Parent Communication Problems. Although language and race had also shown significant main effects on parental CF Knowledge, these variables added less than 5% to the total variance when combined with social class.

Social class was also largely responsible for the significant inverse relationships between parental CF Knowledge and their Index ratings of Information Not Given Before and Doctor-Parent Communication Problems. Together parental social class and their Index ratings of Information Not Given Before predicted almost 60% of the total variance in their CF Knowledge, making them powerful explanatory variables. These findings confirm previous reports of a significant association between parental understanding of the illness and social class membership (Burton 1975 and Nolan, Desmond, Herlich and Hardy 1986).

Paradoxically it would seem that although parents from social classes III, IV and V reported information needs consistently higher than parents from social classes I and II, they received correspondingly less information. This disparity was, in turn, reflected in their lower levels of CF Knowledge. These findings substantiate Pendleton and Bochner's (1980) contention that more explanations are being given to those who need them least. The finding in this study that parents from the lower social classes experienced more communication problems with the doctors, is also well described in the literature (Bochner 1983). Sociolinguistic differences between the social classes (Bernstein 1975) and the diffidence of lower class persons (Pratt, Seligman and Reader 1957; Cartwright 1964; Pendleton and Bochner 1980 and Waitzkin 1984) are the most frequently cited reasons for doctors giving less information to lower class patients. Doctors, for their part, misinterpret the low linguistic competence of lower social class patients and their tendency to ask fewer questions, as reflecting less of a desire for information (Shapiro, Najman, Chang et al 1983) and as showing less of an ability to comprehend (McInlay 1976 and Waitzkin 1984) and recall (Bain 1977) medical matters.

It is important, however, to note that when findings were analyzed separately for fathers and mothers, the effect of social class was not uniform. With the exception of CF Knowledge where the effect of social class was significant for both fathers and mothers, social class was significantly associated only with fathers' Index ratings of Information Needs, Information Not Given Before and Doctor-Parent Communication Problems.

Explanations for the differential social class effect are not entirely clear. Whereas Burton (1975) noted that fathers from the lower social classes were less likely to accompany their wives to clinic and therefore received less information, social class was not significantly associated with fathers' frequency of use of any of the hospital-based sources of information. In addition, Burton (1975) felt that fathers from social classes I and II showed a greater enthusiasm for acquiring more information about CF. This suggestion was also not supported in this study as fathers from the lower social classes expressed information needs greater than those of fathers from the higher social classes.

Burton (1975) felt, furthermore, that fathers from the higher social classes aspired 'to greater mutuality as parents' (p.63) and thus tended to involve themselves more actively in child care responsibilities. Judging from the proportion of fathers, in the present study, who wanted a great deal more information on medical topics, it is equally plausible to surmise that the lack of such information actually prevented lower class fathers from becoming more actively involved in the management of the patients.

One can only speculate that mothers in this study had been more successful in meeting their information needs via the CF literature and the CF Association, both of which were used significantly less frequently by lower class fathers.

Methodologically speaking, the possibility cannot be discounted that the study questionnaire was culturally biased in favour of the higher social classes. Parents from social classes I and II could have been more familiar with the 'test' experience and the format (e.g. multiple choice) of the individual questions

(Messick 1981). Thus their superior CF Knowledge may simply have reflected greater mastery of the test situation. This explanation is, however, unlikely since lower class parents revealed correspondingly higher Information Needs.

Although language and race had a minimal influence on parental CF Knowledge when combined with social class, their independent effects require a brief explanation. The superior CF Knowledge of English speaking parents may have resulted from the scarcity of suitable Afrikaans CF literature. It is also possible that Afrikaans speaking parents misunderstood the information which they had received from predominantly English speaking clinic doctors. Subtle differences in the meaning of questions may also have occurred during the translation process. Since Nolan and colleagues (1986) failed to find a significant language effect in their CF sample, the potential effect of this variable on CF knowledge remains inconclusive.

The significant effect of race on parental CF Knowledge is best explained by the fact that socioeconomic differences paralleled ethnic differences in the sample. The majority (90%) of parents of mixed origin occupied the lower social classes (III, IV and V).

Reasons as to why significantly more parents from social classes III, IV and V expected their child to be cured or very much better in the future are speculative. The significant association between social class and parental Index ratings of Information Not Given Before suggests that lower class parents may, through ignorance, have expected considerable improvement in the patient's future health. Since parents from the lower social classes experienced more problems in communicating with doctors, they may also have found it more difficult to initiate discussion about a sensitive topic such as prognosis.

Given its importance in relation to parental findings, the lack of a significant social class effect on equivalent measures for patients and siblings can only be explained by small sample size.

Collectively these findings provide convincing evidence that social class is an important factor determining how much parents know about CF, what they still wish to know about the illness, whether they have been given information on CF before and, to a lesser extent, whether they have experienced problems in communicating with doctors. These findings can leave no doubt that the medical team will need to pay far more attention to providing information to parents, and more especially fathers, from the lower socioeconomic groups.

Against the above evidence, it was particularly noteworthy that Information Preferences were independent of social class for all family members. The health care team cannot therefore assume that because lower class parents are less forceful in making their information needs known (Burton 1975), that they necessarily have less of a desire to be kept fully informed. It does, however, imply that the onus may well lie with the medical team to identify and meet these wishes.

5.7.5 Shwachman Score, Period Since Diagnosis, Days in Hospital and Number of Clinic Attendances

With one exception, no medical variables were significantly correlated with either CF Knowledge or the measures of Information.

Unlike Nolan and co-workers (1986) who reported a negative association between parental CF knowledge and Shwachman score, no such relationship was noted in this study. The disparity may have resulted from different measures of CF knowledge.

Shwachman Score was significantly associated with only one specific item of information. Where patients were more severely affected by the illness (i.e. obtained lower Shwachman Scores), parents were significantly more likely to have been given information on How to use a nebulizer. As it is clinic policy at this Hospital to prescribe aerosol antibiotic treatment in the presence of permanent pseudomonas colonization (itself an indicator of severity), this finding was not surprising. This finding was also in keeping with the significant relationship noted between age and How to use a nebulizer.

It is noteworthy that Shwachman score was not significantly associated with family members' perception of the patient's Present and Future Health. Similar discrepancies between clinical ratings of the patient's health and those made by the parent or patient have been noted in previous CF research (Frydman 1981 and Strauss and Wellisch 1981). A health versus an illness orientation towards the patient's condition has already been offered as a likely explanation for these findings (Section 5.3).

The lack of a significant relationship between Period since diagnosis and CF Knowledge confirmed similar findings from samples of juvenile diabetic patients (Collier and Etzwiler 1971 and Johnson, Pollak, Silverstein et al 1982) and parents whose children suffered from a variety of paediatric illnesses (Abdurrahman 1978).

Days in hospital was not significantly related to parental CF Knowledge or their ratings on the Information measures. Abdurrahman (1978) had also failed to obtain a significant correlation between parental knowledge and number of hospitalizations. Days in hospital showed a highly significant relationship with the CF Knowledge of male patients only (r_s 0.81, $p = 0.0008$). Given the small number of patients ($n = 9$) involved in this result, it can only be considered a chance finding particularly as the p value was not even suggestive of a relationship for female patients.

Number of clinic attendances was not significantly associated with either mothers' or patients' CF Knowledge or their ratings on the Information measures. Since Index ratings of Frequency of Use of Sources of Informational Support were also not significantly associated with parents' and patients' CF Knowledge, it would seem that the quantity of contact with the medical team may be less important than the quality, in influencing knowledge about CF. (Parents' CF Knowledge, it will be remembered, showed a weak negative relationship with their Index ratings of Doctor-Parent Communication Problems: r_s -0.26, p = 0.005).

The lack of significant correlations in this study, together with the combined evidence from research among other paediatric illnesses, suggest that no medical variables are consistently able to predict disease-related knowledge. Nor is it possible to reliably predict CF family members' information preferences, their information needs or the main sources of their CF information from a patient's medical characteristics. The findings suggest, futhermore, that a patient's clinical status cannot be used as grounds for withholding information. Regardless of degree of severity family members, it seems, want to be kept fully informed about CF.

5.8 RELATIONSHIPS BETWEEN CF KNOWLEDGE, INFORMATION NEEDS AND PERSONAL AND FAMILY ADJUSTMENT

A final aim of the study was to examine the relationship between family members' CF Knowledge, their Information Needs and their self-reported ratings of personal and family adjustment. The stresses which accompany chronic paediatric illness are numerous and have been well-described by Chesler and Yoak (1984). More specifically learning about the illness, its treatment, long-term management and prognosis has been posited as a significant coping task facing patients with a life-threatening illness and their families. Relevant and adequate knowledge about the illness can, on the other hand, equip family members with the cognitive and practical skills needed to fully understand and manage the condition (Johnson 1984).

Inasmuch as the Index ratings of Information Needs reflect a perceived lack of information in family members, these ratings can be conceptualized as a potential source of stress, while their CF Knowledge can be viewed as a potential coping resource. According to the coping model one would therefore expect Information Needs to be positively associated with emotional distress and negatively associated with positive mental health, whereas the converse would be expected between CF Knowledge and adjustment. The direction of the correlations found between these sets of variables supported both these hypotheses although not always at a statistically significant level.

Highly significant positive and negative correlations were noted respectively between self-reported parental Depression and their Information Needs ($r_s = 0.42$, $p = 0.0001$) and their CF Knowledge ($r_s = -0.33$, $p = 0.0003$). A weaker and less significant positive correlation was noted between parental Anxiety and their Information Needs ($r_s = 0.21$, $p < 0.005$). In the case of patients and siblings, only their Information Needs were positively related to their self-rated Anxiety and Depression. Because of low significance levels and small sample sizes, these latter findings require replication. Only parental ratings of Family Functioning showed a weak, positive correlation with their CF Knowledge (r_s 0.22, $p = \ < 0.01$) suggesting that Knowledge of the illness may play a minor role in facilitating Family Functioning.

These findings support the limited available evidence which has linked knowledge of an illness with positive psychological and psychosocial functioning (Klein and Nimorwicz 1982 and Pinkerton, Trauer, Duncan et al 1985).

With respect to the adjustment ratings per se, these were noteworthy for their general absence of personal and family psychopathology, confirming previous empirical work which found that maladjustment and increased stress were not an inevitable consequence of the presence of CF (Smith, Gad and O'Grady 1983; Simmons, Corey, Cowen et al 1985; Cowen, Mok, Corey et al 1986; Holroyd and Guthrie 1986 and Walker, Ford and Donald 1987).

The findings that CF parents, and to a lesser extent patients and siblings, who have higher Information Needs and lower levels of CF Knowledge also experience correspondingly higher levels of depression and anxiety, have important implications for information-giving. Thus, far be it from increasing anxiety and depression by providing too much information, these findings confirm the body of opinion which feels that anxiety and depression are increased by withholding information from patients and their families (Vernick and Karon 1965; Waechter 1971; Jacobs 1971; Spinetta and Maloney 1975, 1978 and Reynolds 1978).

Because correlations cannot establish causality, the possibility cannot be discounted that the more anxious and depressed CF family members were not seeking further information, possibly through a fear of what they might find out. This latter view is less plausible since the majority of family members wanted to be told everything about the condition, irrespective of the nature of this information. Empirical research has, moreover, shown that coping in depressed persons is characterized by seeking rather than avoiding information (Coyne, Aldwin and Lazarus 1981 and Beckham and Adams 1984).

In similar vein, it is not possible to unequivocally interpret the weak positive correlation between parental CF Knowledge and their Family APGAR scores. On the one hand it is reasonable to speculate that increased knowledge of CF contributes to better family functioning (Burton 1975). Yet it is also possible that highly functional families are a more effective medium for the dissemination of disease-related information.

The other finding of practical significance was the existence of a weak positive relationship between family members' Optimism ratings and their CF Knowledge. This association suggests that knowledge of a disease and hope for the future can co-exist. Indeed, similar findings led Cassileth and his colleagues (1980) to conclude that far be it from increasing depression, information 'actually assists many patients in sustaining hopeful attitudes' (p.835).

It must, however, be remembered that this area of research is still in its infancy and definite conclusions regarding the extent and direction of relationships between information needs, disease-related knowledge and adjustment variables await replication in future research particularly of a longitudinal nature.

The combined evidence in this section points to a policy of keeping patients and their families well-informed about all aspects of CF. Psychological well-being is also likely to be promoted as such a policy would be in keeping with family members' expressed preferences towards knowing everything about the illness. To the extent that Koocher (1979) has identified 'exclusionary protectionism' (p.147) (i.e. the withholding of information from patients for their own 'protection') as a significant source of stress for the medical team who have to be on their guard not to let information slip, such a policy could also have direct benefits for the caregivers themselves.

A summary of major findings and recommendations for future research are presented in the final chapter.

CHAPTER 6

SUMMARY AND RECOMMENDATIONS FOR FUTURE RESEARCH

6.1 SUMMARY OF MAJOR FINDINGS AND RECOMMENDATIONS FOR FUTURE RESEARCH

A cross-sectional survey was conducted among a sample of 60 families attending the CF clinic at the Red Cross War Memorial Children's Hospital. Data were collected by means of structured questionnaires from 60 mothers, 54 fathers, 18 patients and 29 siblings. The main goals of the study were to assess family members' medical knowledge of CF, how much they wanted to be told about the illness, their information needs, the main sources of their information and to identify the major sociodemographic and medical correlates of these measures. The study also sought to establish whether a knowledge of CF facilitated coping and conversely whether a lack of information increased psychological distress.

A 63 item CF Knowledge Test, with acceptable psychometric properties, was used to assess medical knowledge. Parents and patients correctly answered almost three-quarters of the items. They obtained scores of over 80% in the sub-sections of general CF knowledge, physiotherapy, GIT symptomatology and treatment and anatomy. They were less well-informed about respiratory symptomatology and treatment, nutrition and genetics. Patients, in particular, performed poorly on genetics items. Parents and patients were least knowledgeable about general medical facts and medical terminology. The panel of CF doctors had overestimated parental knowledge by approximately 24 per cent.

Siblings successfully answered two-thirds of the items on the CF Knowledge Test. Their knowledge of CF was significantly lower than that of parents and patients. Siblings were most knowledgeable about anatomy, GIT symptomatology and treatment, general CF facts and physiotherapy. Their scores ranged between 70% and 80% on these sub-sections. They correctly answered approximately two-thirds of the questions relating to respiratory symptomatology and treatment and nutrition but performed poorly on the sub-sections concerned with genetics, general medical knowledge and terminology.

The following misconceptions and misinformation were shared by a significant proportion of family members: asthma and allergies seldom occur in CF patients, CF patients are more susceptible to the common cold than their siblings, cough mixtures are extremely useful in drying up excess mucus in CF, common colds are caused by bacteria, antibiotics are an effective treatment for viral chest infections, physiotherapy is best given after meals, nebulized antibiotics are most profitably administered before physiotherapy and fats should be excluded from the CF patient's diet.

Many family members had difficulty in applying basic nutritional knowledge. For example, knowing that calories measured the energy content of food did not automatically translate into practical knowledge of what foods a patient should eat to increase energy or weight or to decrease fat consumption. Genetics, on the whole, were poorly understood. Parents, but more especially patients and siblings, had difficulty in grasping concepts of probability. Many family members were also poorly informed about the nature and reliability of the prenatal diagnosis of CF. Knowledge of the medical terms for commonly occurring CF symptoms was absent in well over half the sample of parents and patients and in over 70% of the siblings.

Gender and marital status were associated with performance on the CF Knowledge Test. Mothers and female siblings were significantly better informed than fathers and male siblings. Female patients, too, obtained higher scores than male patients but the difference was not significant. Married mothers were significantly better informed about CF than single mothers. Because the sample of single parents comprised only 6 mothers, marital status was excluded from remaining analyses.

Other significant sociodemographic predictors of parental CF Knowledge were language, race and social class, each of which respectively determined 8.4%, 25.32% and 53.47% of the total variance in parental performance. Parents who were English speaking, of Caucasian origin and came from social classes I and II had significantly more CF Knowledge than parents who were Afrikaans speaking, of Mixed origin and who came from social classes III, IV and V. The combined effect of these variables was responsible for 58.44% of the total variance in parental Test performance. Of these variables social class was the most powerful predictor of parental knowledge. No interaction effects were noted between these variables. No medical variables were significantly associated with CF Knowledge for any family grouping.

No lines x 9

2

In addition to an assessment of family members' factual knowledge of CF, this study also obtained their subjective opinions of the patient's present and future health. Two single item measures were designed for this purpose. Ratings of both Present and Future Health were highly positive. In contrast, the open-ended comments of approximately 40% of fathers and 30% of mothers and siblings revealed obvious anxieties centered on the terminal nature of the illness. Only one patient indicated that he was afraid he might die from his illness. These conflicting findings suggested that an objective rating may not be a reliable measure of family members' perception of a patient's prognosis. With the exception of social class, no other medical or sociodemographic variables were associated with perceptions of Present and Future Health. Significantly more parents from social classes III, IV and V believed that the patient would be cured or very much better in the future.

A 3 item Index of Information Preferences revealed that most family members wanted complete and comprehensive information about the patient's condition. Information Preferences were independent of the effects of sociodemographic and medical variables.

Measures of family members' Information Needs revealed that, in general, they wanted less information about the symptomatology, treatment and genetics of CF and more information about the psychosocial and future implications of the illness. This included a strongly expressed need for more information about the possible effects of CF on the patient's career, social life, marriage and reproductive capacity; and among siblings, a need for more information about carrier status. Patients and siblings were less interested than parents in obtaining much more information on how to cope with feelings aroused by the illness. New information about CF from elsewhere in the world was the most sought after item of information among parents and patients. Among siblings, the most needed area of information related to the different kinds of treatment for CF.

According to Index ratings, fathers revealed the highest Information Needs and patients the lowest. Fathers and siblings required more medical information than mothers and patients. In general, family members had been given far more medical than psychosocial information about CF.

Social class was the most significant predictor of parental Index ratings of their Information Needs and Information Not Given Before. Social class was responsible for 15.84% and 22.38% of the total variance in parental performance on these two Indexes respectively. Parents from social classes I and II were significantly more likely to have been given information about CF and had significantly lower information needs than parents from social classes III, IV and V. The combined effect of social class and parental Index ratings of Information Not Given Before was able to predict almost 60% of the total variance in their scores on the CF Knowledge Test.

With the exception of a relationship between Shwachman score and the increased likelihood that parents had been given information on How to use a nebulizer, no other medical variables correlated significantly with the Index or specific measures of Information Needs and Information Not Given Before for any family grouping.

Parents and patients also rated selected sources of information according to frequency of use and degree of perceived helpfulness. Parents and patients relied most often upon the clinic doctors, their spouse/Mom/Dad, prayer and faith and the CF literature for information and guidance about CF. These sources were also rated as providing the most helpful information and guidance. Fathers, patients and siblings all depended most upon mothers for their information. Mothers, in turn, obtained most of their information about CF from the clinic doctors. Whereas parents considered the CF Association and other parents with a CF child to be moderately helpful sources of information, patients received little helpful information from the Association or other CF patients. The least frequently used sources and those perceived as providing very little helpful information about CF included hospital physiotherapists, the clinic social worker, the dietician, the family doctor, the priest or minister and the local pharmacist.

Social class was significantly associated with how often parents used 3 sources of information. Significantly more fathers from social classes III, IV and V had not sought information from the CF literature and the CF Association, while significantly fewer mothers from these socioeconomic groups had approached the local pharmacist for information about CF.

CF Knowledge was, in turn, significantly associated with the frequency of use of 2 information sources. Fathers and mothers who had had no contact with the CF Association obtained significantly lower scores on the CF Knowledge Test than parents with frequent or occasional contact with this source. Similarly, more frequent use of the CF literature was associated with higher Test scores among fathers.

Based on their Index ratings of Doctor-Parent/Patient Communication problems, the majority of parents and patients did not experience difficulties when seeking CF information from the clinic doctors. Among a minority of parents, however, the tendency to forget what they were told, a lack of privacy at the clinic and the use of jargon ('big words') presented problems.

Parents from social class V experienced significantly more problems when communicating with doctors than parents from social classes I and II. Parental Index ratings of Doctor-Parent Communication Problems were inversely related to their CF Knowledge. The combined effect of Communication Problems and social class were responsible for 25.81% and 53.58% of the total variance in parental Index ratings of Information Not Given Before and parental CF Knowledge respectively.

Social class was a powerful explanatory variable in this study. Among parents it was the most significant predictor of whether they experienced problems when seeking information from doctors, of whether they had been given information about CF, of the magnitude of their information needs and ultimately of how much they knew about the illness. From a coping perspective these findings suggest that parents from social classes I and II had a definite coping advantage over parents from the lower social classes. Parents from the higher social classes experienced less discomfort in the presence of doctors, were more likely to have received the information which they sought and thus had fewer information needs and higher levels of CF knowledge. Small sample numbers may have been responsible for the lack of a significant social class effect for patients and siblings on equivalent measures.

A coping framework was also used to examine the relationship between respondents' CF Knowledge, their Information Needs and their personal and family adjustment. Parents with more medical knowledge about CF and lower Information Needs reported less psychological distress, operationalized in terms of self-reported anxiety and depression. These findings supported the contention that being well informed about an illness would facilitate rather than inhibit coping.

Finally it is necessary to consider these findings against the following shortcomings of the study:

a. The fact that the study was confined to one institution and to a relatively small sample of CF families, limits the generalizability of the results. Certainly the findings for the patients and the siblings remain extremely tentative.

b. Being a cross-sectional investigation, it was not always possible to establish causality when associations between variables were significant. Since it would be illogical to claim that parental CF Knowledge or their Information Needs 'caused' their social class, it is safe to assume that social class was, indeed, a highly significant predictor of their performance on the major dependent variables.

c. For ethical reasons direct questions relating to knowledge of sterility and prognosis were excluded. Furthermore, the single item measure of Perception of Future Health (i.e. knowledge of prognosis) failed to discriminate between respondents.

Given that this study was conceived as an exploratory effort into relatively new areas of CF research, the present data despite their shortcomings have provided an empirical baseline for future research. Clearly most of the findings need replication among larger samples and in other clinics. Against this background, the following suggestions are made for future research:

1. In order to build a body of comparable data, it will be necessary to repeat studies in a standardized manner using equivalent methodologies and measures.
2. In addition to assessing recall of knowledge, future research needs to examine the application of medical knowledge. Such assessments will need to be as close as possible to the kinds of behaviour expected, and the problems encountered, in daily living. Questions might include the following:

   Two hours after a large breakfast you suddenly realize that you forgot to take your enzymes, would you:

Take the required number of enzymes immediately

Take double the amount of enzymes at lunch time

Not do anything and hope that you don't forget again

During a school camp you can't find a secluded place to perform your physiotherapy, would you:

Stay inside and read so as to prevent any bouts of coughing

Exercise more with your friends so as to loosen any secretions

Ask the camp leader if you can return home

While these questions may not have a single solution, patients' responses would provide insight into how they are managing actual situations. Patient and parent education about the illness could, in turn, focus on both supplementing factual knowledge and providing the behavioural skills necessary to cope with new demands.

3. Future research needs to examine the effect of CF Knowledge on compliance.
4. Research needs to examine specific issues relating to the communication of sensitive CF information. Pertinent questions might include: Are doctors adequately trained to provide such information? How much do parents feel that their children should be told about CF? Are parents able to raise sensitive topics in the family, do they feel comfortable in doing so or would they prefer the doctors to provide this information to patients and siblings?

In conclusion, this study has established what and how much a sample of CF families know about CF, it has identified gaps and misconceptions in this knowledge and it has underscored their need for a great deal more information on the psychosocial and future implications of the illness.

For their part, the health care team will need to evaluate family members' understanding of CF at regular intervals. New information can then be provided as patients grow older and needs change, previous explanations can be reinforced and any misunderstandings corrected. Medical educators, in turn, will need to equip doctors with the skills necessary to impart information to chronically ill patients and their families.

APPENDIX 1

PARENTAL QUESTIONNAIRE

CYSTIC FIBROSIS INFORMATION STUDY

It is not always possible for us at the Hospital to know what kinds of information about CF you, as parents, are wanting. In the questions that follow I, therefore, hope to find out what you already know about CF, as well as some of the problems you experience while obtaining information and advice.

Because the questionnaire is quite long, I have tried to make it as easy as possible to answer. Many of the questions simply require a circle, a tick or a short answer. Should you find that the space provided for your answers is not enough to fully explain your opinions and feelings, there is a section for additional comments on the last page.

Please take your time answering the questionnaire.
I would rather you did not discuss your answers with the rest of your family until you have fully answered all the questions. It is YOUR answers and opinions that I want.

Everything that you tell me will be completely confidential, so please be as frank and honest as possible.

I hope you find the questionnaire interesting and enjoyable to answer (in spite of it's length). I am certainly looking forward to receiving your replies.

SECTION 1

The first section of the questionnaire contains questions on the MEDICAL aspects of CF and its treatment.
Should you come across any questions which you cannot answer, simply circle the 'I DON'T KNOW' response.

Please don't look up any answers in books or pamphlets because I'm interested in your immediate responses.

PLEASE ANSWER ALL THE QUESTIONS IN THIS SECTION
BY PLACING A CIRCLE AROUND THE ANSWER YOU CHOOSE

HERE IS AN EXAMPLE:

THE LUNGS ARE PART OF THE:

Respiratory system	[✓ 1]
Digestive system	2
I don't know	3

RESPIRATORY SYSTEM is the correct answer, so you would place your circle around Number One.

REMEMBER THIS IS NOT A TEST!

IT IS ONLY TO HELP ME FIND OUT
WHAT YOU UNDERSTAND ABOUT CF

1 CYSTIC FIBROSIS IS AN ILLNESS WHICH AFFECTS THE

Lungs and pancreas	1
Liver and stomach	2
I don't know	3

2 IN CF, MUCOUS SECRETIONS ARE A PROBLEM BECAUSE THEY ARE

The wrong colour	1
Thick and sticky	2
I don't know	3

3 THE MOST ACCURATE WAY TO DIAGNOSE CF IS BY

A chest X-ray	1
A sweat test	2
I don't know	3

4 THE CAUSE OF CYSTIC FIBROSIS IS

Inheritance	1
Bad lungs	2
I don't know	3

5 CF CHILDREN MAY SUFFER FROM FREQUENT CHEST INFECTIONS BECAUSE

They lack vitamin C	1
Germs become trapped in the mucus in their lungs	2
I don't know	3

6 DURING A CHEST INFECTION, THE AMOUNT OF MUCUS IN THE LUNGS OF CF CHILDREN IS LIKELY TO

Increase	1
Decrease	2
I don't know	3

7 A CF CHILD WHO HAS EXCESSIVE MUCUS IN HIS LUNGS IS LIKELY TO HAVE

An increased appetite	1
A decreased appetite	2
I don't know	3

8 DOCTORS OFTEN REFER TO SHORTNESS OF BREATH AS

Dyspnoea	1
Clubbing	2
I don't know	3

9 THE MEDICAL NAME FOR COUGHING UP BLOOD IS

Bronchospasm	1
Haemoptysis	2
I don't know	3

10 ASTHMA AND ALLERGIES

Often occur in children with CF	1
Are seldom found in children with CF	2
I don't know	3

11 COMMON COLDS ARE CAUSED BY

Bacteria	1
Viruses	2
I don't know	3

12 WHICH ONE OF THE FOLLOWING SYMPTOMS SHOWS THAT A CF CHILD PROBABLY HAS A BACTERIAL CHEST INFECTION?

Green sputum	1
Sneezing	2
I don't know	3

13 IN CF, COUGH MIXTURES WHICH DRY UP THE EXCESS MUCUS IN THE LUNGS ARE

Extremely useful	1
To be avoided	2
I don't know	3

14 COMPARED TO THEIR BROTHERS AND SISTERS, CF CHILDREN ARE LIKELY TO CATCH

The same number of common colds	1
A much larger number of common colds	2
I don't know	3

15 A CF CHILD WHO HAS A PERSISTENT COUGH SHOULD BE

Allowed to attend school if he/she is feeling well	1
Kept at home because he/she will infect his/her classmates	2
I don't know	3

16 THE AVERAGE 'NORMAL' TEMPERATURE OF A CHILD, IF TAKEN BY MOUTH, IS

37°C (98.6°F)	1
38°C (100.4°F)	2
I don't know	3

17 A CHILD'S TEMPERATURE IS USUALLY HIGHEST IN THE

Early morning	1
Late afternoon	2
I don't know	3

18 ANTIBIOTICS ARE VERY EFFECTIVE IN THE TREATMENT OF

Bacterial chest infections	1
Viral chest infections	2
I don't know	3

19 IF A CHEST INFECTION IS BEING TREATED WITH ANTIBIOTICS, PHYSIOTHERAPY

Can be stopped altogether	1
Should continue everyday	2
I don't know	3

20 IN CF, THE MAIN REASON FOR DOING CHEST PHYSIOTHERAPY IS TO

Cure the lung infections	1
Remove harmful secretions	2
I don't know	3

21 PHYSIOTHERAPY SHOULD BE PERFORMED

Only when a CF child is coughing a great deal	1
Everyday, even if the CF child is well	2
I don't know	3

22 IN CF, CHEST PHYSIOTHERAPY LOOSENS THE MUCUS SO THAT IT CAN BE

Swallowed	1
Coughed up	2
I don't know	3

23 OUTDOOR EXERCISE AND SPORT FOR CF CHILDREN SHOULD BE

Avoided because they cause too much coughing	1
Encouraged because they loosen the mucus	2
I don't know	3

24 SMALL CF CHILDREN SHOULD RECEIVE THEIR PHYSIOTHERAPY

Before meals	1
After meals	2
I don't know	3

25 IN OLDER CF CHILDREN, THE MOST EFFECTIVE TIME TO PERFORM PHYSIOTHERAPY IS

In the morning before breakfast	1
Immediately after breakfast before going to school	2
I don't know	3

26 WHEN A CF CHILD HAS A CHEST INFECTION, AN ANTIBIOTIC MAY BE GIVEN THROUGH A NEBULIZER

Before physiotherapy	1
After physiotherapy	2
I don't know	3

27 WHICH OF THE FOLLOWING ARE PARTICULARLY HARMFUL TO THE HEALTH OF A CF CHILD?

Swimming and running	1
Cigarette smoke	2
I don't know	3

28 PANCREATIC ENZYMES ARE SUBSTANCES WHICH

Reduce the acidity in the stomach	1
Speed up the digestion of food	2
I don't know	3

29 CHILDREN WITH CF HAVE

Too few pancreatic enzymes	1
Too many pancreatic enzymes	2
I don't know	3

30 IF A CHILD WITH CF STARTS TO HAVE MORE SMELLY, OILY STOOLS THAN USUAL THE MOST LIKELY REASON WOULD BE

The side-effects of the pancreatic enzymes	1
Too much fat in the diet	2
I don't know	3

31 LARGE, GREASY AND SMELLY STOOLS ARE CALLED

Steatorrhoea	1
Diarrhoea	2
I don't know	3

32 CHILDREN WITH CF SHOULD TAKE PANCREATIC ENZYMES

With ALL their meals and snacks	1
ONLY with their three main meals	2
I don't know	3

33 THE BEST TIME TO TAKE PANCREATIC ENZYMES IS

After meals	1
Before meals	2
I don't know	3

34 PEOPLE WITH CF SHOULD EAT A

Well-balanced diet that the whole family can enjoy	1
Carefully planned fat-free diet	2
I don't know	3

35 CALORIES (OR KILOJOULES) MEASURE THE AMOUNT OF

Energy in our food	1
Vitamins in our food	2
I don't know	3

36 BECAUSE MANY CF CHILDREN HAVE DIFFICULTY DIGESTING FAT, THEY HAVE TO OBTAIN THEIR EXTRA CALORIES FROM

Proteins	1
Carbohydrates	2
I don't know	3

37 CALOREEN IS A USEFUL SOURCE OF

Carbohydrate	1
Fat	2
I don't know	3

38 INSTEAD OF WHOLE (REGULAR) MILK, SOME PARENTS GIVE SKIM MILK TO THEIR CF CHILDREN TO DRINK BECAUSE IT CONTAINS

Less sugar	1
Less fat	2
I don't know	3

39 MOST CF CHILDREN USE UP PLENTY OF ENERGY. WHICH OF THE FOLLOWING SNACKS CONTAINS THE MOST AMOUNT OF ENERGY?

Two raw carrots	1
One medium-sized banana	2
I don't know	3

40 IF THE DOCTOR SUGGESTS THAT A CF CHILD SHOULD EAT LESS FAT, WHICH ONE OF THE FOLLOWING SHOULD BE LEFT OUT OF HIS DIET?

Jam	1
Peanut-butter	2
I don't know	3

41 SPAGHETTI, RICE AND PORRIDGE ARE EXAMPLES OF

Carbohydrates	1
Fats	2
I don't know	3

42 BUTTER AND CHEESE CONTAIN PLENTY OF

Carbohydrates	1
Fat	2
I don't know	3

43 HOW MANY PANCREATIC ENZYMES TO TAKE WITH EACH MEAL DEPENDS ON THE AMOUNT OF

Carbohydrate in the food	1
Fat in the food	2
I don't know	3

44 CHILDREN WITH CF OFTEN NEED TO PUT ON WEIGHT. WHICH OF THE FOLLOWING SNACKS, IF EATEN REGULARLY, IS LIKELY TO LEAD TO THE GREATEST INCREASE IN WEIGHT?

One large apple	1
One slice of fruit cake	2
I don't know	3

45 BECAUSE MANY CHILDREN WITH CF HAVE AN INADEQUATE SUPPLY OF PANCREATIC ENZYMES, THEY SHOULD

Eat as much fat as their bodies can tolerate	1
Exclude fats completely from their diet	2
I don't know	3

46 A WELL-BALANCED, LOW-FAT DIET CAN

Cure the symptoms of CF	1
Reduce the number of oily, smelly stools	2
I don't know	3

47 IN SUMMER, CHILDREN WITH CF MAY SWEAT A GREAT DEAL, IT IS THEREFORE IMPORTANT THAT THEY ADD PLENTY OF

Sugar to their meals	1
Salt to their meals	2
I don't know	3

48 IF AN ILLNESS IS INHERITED, THIS MEANS THAT IT IS

Infectious	1
Runs in families	2
I don't know	3

49 FOR A CHILD TO BE BORN WITH CF, IT IS NECESSARY THAT

Only ONE parent carries the CF gene	1
BOTH parents carry the CF gene	2
I don't know	3

50 IT IS NOW POSSIBLE TO DETECT WHETHER AN UNBORN BABY MIGHT HAVE CF BY MEANS OF

A blood test	1
Amniocentesis	2
I don't know	3

51 THE PRENATAL DIAGNOSIS OF CF IS

One hundred per cent reliable	1
About ninety per cent reliable	2
I don't know	3

52 EACH TIME A COUPLE WHO BOTH CARRY THE CF GENE HAVE A CHILD, THEIR CHANCES OF HAVING A CHILD WITH CF ARE

1 in 2	1
1 in 4	2
I don't know	3

53 WHERE BOTH PARENTS ARE CARRIERS OF THE CF GENE, THE CHANCES OF THEIR HAVING A CHILD WHO DOES NOT HAVE CF ARE

25%	1
75%	2
I don't know	3

54 WHERE A COUPLE ALREADY HAS TWO CHILDREN WITH CF, WHAT ARE THEIR CHANCES OF HAVING A THIRD CHILD WITH THE ILLNESS?

1 in 3	1
1 in 4	2
I don't know	3

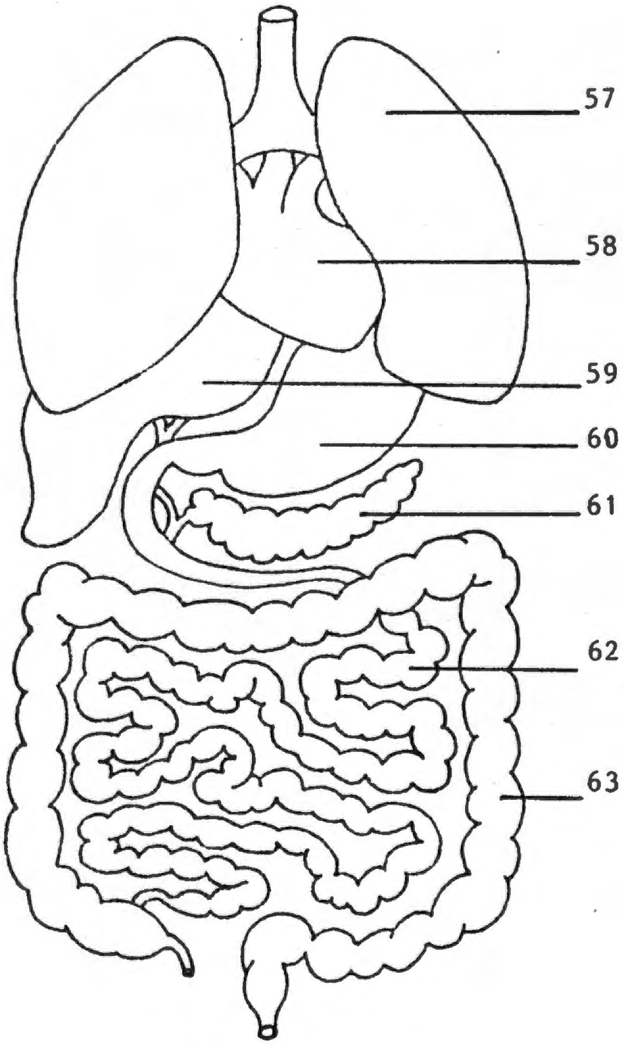
55 PEOPLE WITH CF

Usually outgrow their illness	1
Have a life-long illness	2
I don't know	3

56 WHICH ONE OF THE FOLLOWING IS NOT RECOMMENDED FOR THE OLDER PERSON WITH CF?

Getting married	1
Smoking cigarettes	2
I don't know	3

57-63
FINALLY CAN YOU NAME THE 7 PARTS OF THE BODY THAT YOU SEE BELOW? SIMPLY FILL IN THE NAME OF THE PARTS IN THE BLANK SPACES NEXT TO THE QUESTION NUMBERS.



SECTION 2

THE REST OF THE QUESTIONNAIRE IS CONCERNED WITH YOUR PERSONAL OPINIONS AND FEELINGS SO, FROM NOW ON, THERE WILL BE NO RIGHT OR WRONG ANSWERS.

If you should find that you don't really have an opinion about something, could you nevertheless choose an answer that is closest to how you feel at the moment.

Although you may feel that some questions are similar, there are differences between them and each one has been included to help me understand your problems and feelings a little better.

Learning that your child has CF marks the beginning of an endless task of gathering and understanding large amounts of information about the illness and its treatment. What's more, much of this information seems to be in a foreign language.

Could you please indicate, by placing a tick in the appropriate column, if you would like some more information about the areas listed below. Could you also, please, indicate whether you have ever been given this kind of information before. Place the SECOND tick in either the YES or NO column.

You will notice that I have also included a few 'non-medical' items in the list. Please could you answer ALL the items, even though they may not apply directly to your child at the moment.

You will find an example on the following page.

HERE IS AN EXAMPLE:

HOW MUCH INFORMATION WOULD YOU LIKE ABOUT:	A GREAT DEAL MORE INFORMATION	A LITTLE MORE INFORMATION	I HAVE ENOUGH INFORMATION	I DO NOT WANT TO KNOW THIS	HAVE YOU BEEN GIVEN THIS INFORMATION BEFORE?	
					YES	NO
a. Genetics of CF			✓		✓	
b. How to treat chest infections		✓			✓	

(EACH ITEM HAS TWO TICKS)

	HOW MUCH INFORMATION WOULD YOU LIKE ABOUT:	A GREAT DEAL MORE INFORMATION	A LITTLE MORE INFORMATION	I HAVE ENOUGH INFORMATION	I DO NOT WANT TO KNOW THIS	HAVE YOU BEEN GIVEN THIS INFORMATION BEFORE?	
						YES	NO
1	How the human body works						
2	Symptoms of CF						
3	How CF is inherited						
4	When and how to take antibiotics						
5	When and how to take pancreatic enzymes						
6	How to perform physiotherapy						
7	How to use a nebulizer						
8	Forms of exercise and sport that are suitable for people with CF						
9	Healthy meals for CF children						
10	New information, about CF, from other places in the world						
11	Detailed results of any tests performed on your child when he/she attends clinic e.g. X rays, sputum tests						

	HOW MUCH INFORMATION WOULD YOU LIKE ABOUT:	A GREAT DEAL MORE INFORMATION	A LITTLE MORE INFORMATION	I HAVE ENOUGH INFORMATION	I DO NOT WANT TO KNOW THIS	HAVE YOU BEEN GIVEN THIS INFORMATION BEFORE?	
						YES	NO
12	Possible complications of CF						
13	How to deal with your child's feelings about having CF e.g. feelings of anger, sadness and frustration						
14	What to tell your child about his/her illness						
15	What to tell your other children about CF						
16	How CF might affect your child's career, social life and marriage						
17	How CF might affect your child's chances of having his/her own children						
18	How serious your child's illness is						
19	What to expect if your child's illness gets worse						
20	How to answer your child's questions about whether he might die from his illness						

(DID YOU REMEMBER TO TICK EACH ITEM TWICE?)

Are there any other areas in which you would like more information and advice about CF? If so, please list them in the space below:

____

____

____

____

Parents differ as to HOW MUCH information they would like about their child's condition. Would you please indicate to what extent you would agree or disagree with the statements below. Place a tick in the column you choose. (ONE tick only this time)

	STRONGLY AGREE	AGREE	NEITHER AGREE OR DISAGREE	DISAGREE	STRONGLY DISAGREE
1 It is my right to know everything about my child's condition					
2 Knowing all the facts about CF will only make me miserable					
3 I want as much information as possible about CF - good orbad					

The following questions are concerned with HOW you obtain information about CF.

There are many different WAYS of learning more about CF. In the past twelve months or so, HOW OFTEN have you tried to obtain information and guidance about your child's illness from the sources listed below? Place a tick in the appropriate column for each item.

		MANY TIMES	OCCASIONALLY	NOT AT ALL
1	Other parents who also have a child with CF			
2	Your family doctor			
3	The doctors at the CF clinic			
4	The physiotherapists at the hospital			
5	The social worker at the CF clinic			
6	The dietician at the hospital			
7	Books, pamphlets and magazines about CF			
8	The CF Association			
9	Prayer and Faith			
10	Your priest or minister			
11	Your husband or wife			
12	Your local pharmacy			

In the previous question, you indicated how often you used the above sources when you needed information and advice about CF.

Now I'd like to know HOW MUCH helpful information and guidance you have, in fact, received from these sources. Once again, place a tick in the appropriate column for each item.

		A great deal	Quite a lot	Not very much	None at all
1	Other parents who also have a child with CF				
2	Your family doctor				
3	The doctors at the CF clinic				
4	The physiotherapists at the hospital				
5	The social worker at the CF clinic				
6	The dietician at the hospital				
7	Books, pamphlets and magazines about CF				
8	The CF Association				
9	Prayer and Faith				
10	Your priest or minister				
11	Your husband or wife				
12	Your local pharmacy				

If you have placed any ticks in the last two columns ('not very much' and 'none at all'), please could you explain, IN YOUR OWN WORDS, why these sources have not been helpful to you. For example, you may not have a family doctor or local pharmacist, or you may find pamphlets too difficult to understand etc.

____

____

____

____

____

____

Who, from the above list, is your MOST IMPORTANT SOURCE of information and guidance about CF e.g. the clinic doctors, your husband or wife etc? Only choose ONE source and write the name in the space below.

____

Can you think of any other SOURCES of information about CF, which you have found PARTICULARLY HELPFUL? If so, could you mention your suggestions in the space below.

____

____

____

____

All parents have worries and uncertainties because of their child's illness. Your answers to the statements below will help me to understand YOUR particular concerns and feelings a little better. Simply indicate, with a tick in the appropriate column, whether or not you agree or disagree, with each of the statements below.

		Strongly Agree	Agree	Neither Agree or Disagree	Disagree	Strongly Disagree
1	In uncertain times, I usually hope for the best					
2	I believe in the saying that 'every cloud has a silver lining'					
3	I can really count on the doctors for honest answers about my child's condition					
4	I hardly ever expect good things to happen to me					
5	The doctors will think I'm a nuisance if I ask too many questions about CF					
6	I always look on the bright side of things					
7	If something can go wrong for me, it will					
8	I often forget what the doctors tell me about CF					

	Strongly Agree	Agree	Neither Agree or Disagree	Disagree	Strongly Disagree
9 The doctors don't explain things to me because they think I won't understand					
10 I hardly ever expect things to go my way					
11 I'm always optimistic about the future					
12 The doctors really understand how I feel about my child's illness					
13 The lack of privacy at the clinic makes it difficult to ask the doctors questions					
14 Things never work out the way I want them to					
15 The doctors use too many big words when they explain things to me					
16 The doctors are usually too busy to answer my questions about CF					

How would you describe your CF child's health at present?

Excellent ☐
Very good ☐
Good ☐
Fair ☐
Poor ☐
Very poor ☐

As your CF child gets older, do you expect him/her to

Be cured ☐
Be very much better ☐
Be a little better ☐
Remain the same ☐
Be a little worse ☐
Be a lot worse ☐
A great deal worse ☐

What worries you MOST about your child's illness?

Answer in your own words

____

____

What do you feel is the WORST sickness a child can have?

____

SECTION 3

FINALLY I'D LIKE TO ASK YOU SOME QUESTIONS ABOUT YOURSELF AND YOUR FAMILY.

Firstly, your family:

We know that families can be very different - especially when it comes to how much family members talk to each other and share problems.

Your answers to the next few questions will help me to understand your family a little better.

For each item, place a tick in the column that comes closest to how you feel.

		MOST OF THE TIME	SOME OF THE TIME	HARDLY EVER
1	I am satisfied that I can turn to my family for help, when something is troubling me			
2	I am satisfied with the way our family talks things over and shares problems			
3	I am satisfied that my family accepts and supports my wishes to take on new activities or directions			
4	I am satisfied with the way my family expresses affection and responds to my feelings such as anger, sadness or love.			
5	I am satisfied with the way our family shares time together			

NOW I HAVE SOME QUESTIONS ABOUT HOW YOU HAVE BEEN FEELING LATELY

Read each item and place a tick in the box opposite the reply which comes closest to how you have been feeling in the PAST FEW WEEKS.

Don't take too long over your replies - simply indicate your immediate responses.

(Only ONE tick per item)

I feel tense or 'wound up':

Most of the time ☐
A lot of the time ☐
Time to time, occasionally ☐
Not at all ☐

I still enjoy the things I used to enjoy:

Definitely as much ☐
Not quite so much ☐
Only a little ☐
Hardly at all ☐

I get a sort of frightened feeling as if something awful is about to happen:

Very definitely and quite badly ☐
Yes, but not too badly ☐
A little, but it doesn't worry me ☐
Not at all ☐

I can laugh and see the funny side of things:

As much as I always could ☐
Not quite so much now ☐
Definitely not so much now ☐
Not at all ☐

Worrying thoughts go through my mind:

A great deal of the time ☐
A lot of the time ☐
From time to time but not too often ☐
Only occasionally ☐

I feel cheerful:

Not at all ☐
Not often ☐
Sometimes ☐
Most of the time ☐

I can sit at ease and feel relaxed:

Definitely ☐
Usually ☐
Not often ☐
Not at all ☐

I feel as if I am slowed down:

Nearly all the time ☐
Very often ☐
Sometimes ☐
Not at all ☐

I get a sort of frightened feeling like 'butterflies' in the stomach:

Not at all ☐
Occasionally ☐
Quite often ☐
Very often ☐

I have lost interest in my appearance:

Definitely ☐
I don't take so much care as I should ☐
I may not take quite as much care ☐
I take just as much care as ever ☐

I feel restless as if I have to be on the move:

Very much indeed ☐
Quite a lot ☐
Not very much ☐
Not at all ☐

I look forward with enjoyment to things:

As much as ever I did ☐
Rather less than I used to ☐
Definitely less than I used to ☐
Hardly at all ☐

I get sudden feelings of panic:

Very often indeed ☐
Quite often ☐
Not very often ☐
Not at all ☐

I can enjoy a good book or radio or TV programme:

Often ☐
Sometimes ☐
Not often ☐
Very seldom ☐

IF YOU HAVE ANYTHING ELSE TO ADD, THERE IS PLENTY OF SPACE ON THE NEXT PAGE. I WOULD REALLY APPRECIATE YOUR COMMENTS AND SUGGESTIONS

ADDITIONAL COMMENTS AND SUGGESTIONS:

If your family would like a summary of the findings of the CF INFORMATION STUDY, would you please place a tick in the appropriate box.

YES ☐
NO ☐

THANK YOU VERY MUCH FOR COMPLETING THE QUESTIONNAIRE. Without your help this study would'nt have been possible.

I shall call to collect your questionnaires within a few days, at a time which is convenient to your family. In the meantime, please could you put your questionnaire into the envelope and seal it.

APPENDIX 2

PATIENTS' QUESTIONNAIRE

CYSTIC FIBROSIS INFORMATION STUDY

How much do you know about your illness? Who told you about CF? Do you have any questions you would like to ask about CF? These are just a few of the things I'm going to ask you in this questionnaire. Your answers will help me to better understand how you feel about CF.

Because the questionnaire is quite long, I have tried to make it as simple as possible for you to answer. In fact, MOST of the questions only need a circle, a tick or a short answer. Should you have anything else to add, there is a special section for 'additional comments' on the last page.

Please take your time answering the questionnaire.
I would rather you didn't discuss your answers with the rest of your family until you have finished answering all the questions - it's YOUR answers and feelings that I am looking for.

Nobody else will see your answers so you can say exactly how you feel.

I hope you enjoy answering the questionnaire. I'm certainly looking forward to your replies.

SECTION 1

Refer pages 2 - 11 of Parental Questionnaire (Appendix 1)

SECTION 2

FROM NOW ON, THERE WILL BE NO RIGHT OR WRONG ANSWERS TO THE QUESTIONS. INSTEAD, I WANT TO FIND OUT YOUR FEELINGS AND OPINIONS.

In order to take proper care of yourself, you need to know a lot about your illness and its treatment.

Let's begin by seeing what YOU would like to know about your illness.

Please indicate, by placing a tick in the appropriate column, if you would like to know more about the areas I have listed below. You will notice that I have included both medical and non-medical items.

Could you please also indicate whether you have ever been given this kind of information before. Place the second tick in either the 'yes' or 'no' column.

You will find an example on the next page.

HERE IS AN EXAMPLE:

HOW MUCH INFORMATION WOULD YOU LIKE ABOUT:	A GREAT DEAL MORE INFORMATION	A LITTLE MORE INFORMATION	I HAVE ENOUGH INFORMATION	I DO NOT WANT TO KNOW THIS	HAVE YOU BEEN GIVEN THIS INFORMATION BEFORE?	
					YES	NO
a. Genetics of CF	✓					✓
b. How to treat chest infections			✓		✓	

(EACH ITEM HAS TWO TICKS)

	HOW MUCH INFORMATION WOULD YOU LIKE ABOUT:	A GREAT DEAL MORE INFORMATION	A LITTLE MORE INFORMATION	I HAVE ENOUGH INFORMATION	I DO NOT WANT TO KNOW THIS	HAVE YOU BEEN GIVEN THIS INFORMATION BEFORE?	
						YES	NO
1	How the human body works						
2	Symptoms of CF						
3	How CF is inherited						
4	When and how to take your antibiotics						
5	When and how to take your pancreatic enzymes						
6	How to perform your physiotherapy						
7	How to use a nebulizer						
8	Forms of exercise and sport that are suitable for people with CF						
9	What kinds of food to eat to increase your weight						
10	New information, about CF, from other places in the world						
11	Detailed results of your X rays, sputum tests etc.						
12	Possible complications of CF						

	HOW MUCH INFORMATION WOULD YOU LIKE ABOUT:	A GREAT DEAL MORE INFORMATION	A LITTLE MORE INFORMATION	I HAVE ENOUGH INFORMATION	I DO NOT WANT TO KNOW THIS	HAVE YOU BEEN GIVEN THIS INFORMATION BEFORE?	
						YES	NO
13	How to deal with your feelings about having CF e.g. sad, angry, frustrated feelings						
14	What to tell your friends about your illness						
15	How to talk to your parents about your illness						
16	How CF might affect your career, social life and marriage						
17	How serious your illness is						
18	What to expect if your illness gets worse						
19	How other people with CF are coping with their illness						

(DID YOU REMEMBER TO TICK EACH ITEM TWICE?)

Are there any other areas in which you would like more information and advice about your illness? If so, please list them in the space below:

____

____

____

____

People feel differently about HOW MUCH they would like to know about their illness. HOW MUCH DO YOU WANT TO KNOW ABOUT CF? Simply indicate whether you agree or disagree with the statements below. Place a tick in the column which comes closest to describing how you feel. (ONE tick only this time)

	STRONGLY AGREE	AGREE	NEITHER AGREE OR DISAGREE	DISAGREE	STRONGLY DISAGREE
1 It is my right to know everything about my condition					
2 Knowing all the facts about CF will only make me miserable					
3 I want as much information as possible about CF - good or bad					

Would your say your health is:

Tick only one box

Excellent ☐
Very good ☐
Good ☐
Fair ☐
Poor ☐
Very poor ☐

As you get older, do you expect your illness to:

Be cured ☐
Be very much better ☐
Be a little better ☐
Remain the same ☐
Be a little worse ☐
Be a lot worse ☐
Be a great deal worse ☐

What worries YOU most about your illness?

Answer in your own words.

____

____

____

What do you feel is the WORST sickness a person can have?

____

The following questions are concerned with HOW you obtain information about CF.

There are many different WAYS of obtaining information and advice about your illness. In the past twelve months or so, when you have needed information and guidance about CF, HOW OFTEN have you used any of the sources listed below? Place a tick in the appropriate column for each item.

		MANY TIMES	OCCASIONALLY	NOT AT ALL
1	Other people who also have CF			
2	Your family doctor			
3	The doctors at the CF clinic			
4	The physiotherapists at the hospital			
5	The social worker at the CF clinic			
6	The dietician at the hospital			
7.	Books, pamphlets and magazines about CF			
8	The CF Association			
9	Prayer and Faith			
10	Your priest or minister			
11	Your Mom			
12	Your Dad			
13	Your Chemist			

In the previous question, you indicated how often you used the above sources, when you needed information and advice about your illness.

Now I'd like to know HOW MUCH HELPFUL information and guidance you have, in fact, received from these sources. Once again, place a tick in the appropriate column for each item.

		A great deal	Quite a lot	Not very much	None at all
1	Other people who also have CF				
2	Your family doctor				
3	The doctors at the CF clinic				
4	The physiotherapists at the hospital				
5	The social worker at the CF clinic				
6	The dietician at the hospital				
7	Books, pamphlets and magazines about CF				
8	The CF Association				
9	Prayer and Faith				
10	Your priest or minister				
11	Your Mom				
12	Your Dad				
13	Your chemist				

If you have placed any ticks in the last two columns ('not very much' and 'none at all'), please could you explain, IN YOUR OWN WORDS, why these sources have not been helpful to you. For example, you may not have a family doctor or you may never go to a chemist, or you may find pamphlets too difficult to understand etc.

Who, from the above list, is your MOST IMPORTANT SOURCE of information and guidance about CF e.g. the clinic doctors, your Mom or Dad etc? Only choose ONE source and write the name in the space below.

Can you think of any other SOURCES of information about CF, which you have found PARTICULARLY HELPFUL? If so, could you mention your suggestions in the space below.

____

____

____

____

Do you have an opportunity to talk to the doctors alone when you attend the CF clinic?

Tick one box

YES ☐

NO ☐

Do you prefer to have your Mom or Dad present when you talk to the clinic doctors?

YES ☐

NO ☐

How accurately do the following statements describe you? Simply indicate how much you agree or disagree with each item, by putting a tick in the column which comes closest to how you feel. Remember that there are no right or wrong answers - I only want your honest opinions. Although some of the statements may seem similar, there are differences between them and each one has been included to help me understand how you feel.

		Strongly Agree	Agree	Neither Agree or Disagree	Disagree	Strongly Disagree
1	In uncertain times, I usually hope for the best					
2	I believe in the saying that 'every cloud has a silver lining'					
3	I can really count on the doctors for honest answers about my condition					
4	I hardly ever expect good things to happen to me					
5	The doctors will think I'm a nuisance if I ask too many questions about CF					
6	I always look on the bright side of things					
7	If something can go wrong for me, it will					
8	I often forget what the doctors tell me about CF					
9	The doctors don't explain things to me because they think I won't understand					
10	I hardly ever expect things to go my way					

		Strongly Agree	Agree	Neither Agree or Disagree	Disagree	Strongly Disagree
11	I'm always optimistic about the future					
12	The doctors really understand how I feel about my illness					
13	The lack of privacy at the clinic makes it difficult to ask the doctors questions					
14	Things never work out the way I want them to					
15	The doctors use too many big words when they explain things to me					
16	The doctors are usually too busy to answer my questions about CF					

I'M SURE THAT YOU ARE WONDERING IF THIS QUESTIONNAIRE WILL EVER COME TO AN END. I KNOW IT IS VERY LONG - BUT YOU ARE ALREADY HALF-WAY THROUGH IT.

SECTION 3

FINALLY I'D LIKE TO ASK YOU SOME QUESTIONS ABOUT YOURSELF AND YOUR FAMILY.

Firstly, I'd like to ask you a few questions about your family:

We know that families can be very different - especially when it comes to how much family members talk to each other and share problems.

Your answers to the next few questions will help me to understand your family a little better.

For each item, place a tick in the column that comes closest to how you feel.

		MOST OF THE TIME	SOME OF THE TIME	HARDLY EVER
1	I am satisfied that I can turn to my family for help when something is troubling me			
2	I am satisfied with the way our family talks things over and shares problems			
3	I am satisfied that my family accepts and supports my wishes to take on new activities or directions			
4	I am satisfied with the way my family expresses affection and responds to my feelings such as anger, sadness or love			
5	I am satisfied with the way our family shares time together			

You're coming very close to the end of the questionnaire.

The final questions are concerned with your feelings. No two people with CF are ever alike - each of you thinks and feels differently about things in your life. Your responses to the following statements will help me to understand YOU a little better.

For each item, place a tick in the column which comes closest to how you have been feeling in the PAST TWO WEEKS.

		MOST OF THE TIME	SOME OF THE TIME	NEVER
1	I still look forward to things as much as I used to			
2	I sleep very well			
3	I feel like crying			
4	I like to have fun			
5	I feel like running away			
6	I get tummy aches			
7	I have lots of energy			
8	I enjoy my food			
9	I can stick up for myself			
10	I feel my life isn't worth living			
11	I am good at the things I do			
12	I enjoy the things I do			
13	I like talking to my family			
14	I have horrible dreams			
15	I feel very lonely			
16	I am easily cheered up			
17	I feel so sad I can hardly stand it			
18	I feel very bored			

Once more could you please indicate whether or not the following statements describe how you feel. Read each one carefully and put a tick in the YES column if you think a statement is true for you. If it does not apply to you, put your tick in the NO column.

		YES this is true for me	NO this is not true for me
1	I have trouble making up my mind		
2	I get nervous when things don't go right for me		
3	Other people seem to do things more easily than I can		
4	I often have trouble getting my breath		
5	I worry a lot of the time		
6	I am afraid of a lot of things		
7	I get angry easily		
8	I worry about what my parents will say		
9	I feel that other people do not like the way I do things		
10	It's hard for me to get to sleep at night		
11	I worry about what other people think of me		
12	I feel alone even when there are people with me		
13	I often have a frightened feeling like 'butterflies' in my tummy		
14	My feelings get hurt easily		
15	My hands often feel sweaty		
16	I am tired a lot		

		YES this is true for me	NO this is not true for me
17	I worry about what is going to happen		
18	Other children/people are happier than I am		
19	I have bad dreams		
20	My feelings get hurt easily when I am moaned at		
21	I feel that people will tell me I do things the wrong way		
22	I sometimes wake up scared		
23	I worry when I go to bed at night		
24	It is hard for me to keep my mind on my work/school work		
25	I can't sit still for long		
26	I am nervous		
27	A lot of people are against me		
28	I often worry about something awful happening to me		

If you have anything else to add, please use the space below. I would really appreciate your comments and suggestions.

____

____

____

Thank you very much for completing this questionnaire.

APPENDIX 3

SIBLINGS' QUESTIONNAIRE

CYSTIC FIBROSIS INFORMATION STUDY

How much do you know about CF? Would you like to have some more information about CF? Who answers most of your questions about CF? These are the sorts of things I'm going to ask you in this questionnaire. And from your responses I hope to get a better understanding of what you know and feel about your brother or sister's illness.

Because the questionnaire is quite long, I have tried to make it as simple as possible for you to answer. In fact, MOST of the questions only need a circle, a tick or a short answer. Should you have anything else to add, there is a special section for 'additional comments' on the last page.

Please take your time answering the questionnaire.
I would rather you didn't discuss your answers with the rest of your family until you have finished answering all the questions - it's YOUR answers and feelings that I am looking for.

Nobody else will see your answers so you can say exactly how you feel.

I hope you enjoy answering the questionnaire. I'm certainly looking forward to your replies.

SECTION 1

Refer pages 2 - 11 of Parental Questionnaire (Appendix 1)

SECTION 2

FROM NOW ON, THERE WILL BE NO RIGHT OR WRONG ANSWERS TO THE QUESTIONS. INSTEAD, I WANT TO FIND OUT YOUR FEELINGS AND OPINIONS.

Living within a family where one member has cystic fibrosis, means that you will already have learnt quite a lot about the illness. You may, however, still have some areas where you would like some more information.

Please indicate, by placing a tick in the appropriate column, if you would like to know more about the areas I have listed below. You will notice that I have included both medical and non-medical items.

Could you please also indicate whether you have ever been given this kind of information before. Place the second tick in either the 'yes' or 'no' column.

You will find an example on the next page.

HERE IS AN EXAMPLE:

HOW MUCH INFORMATION WOULD YOU LIKE ABOUT:	A GREAT DEAL MORE INFORMATION	A LITTLE MORE INFORMATION	I HAVE ENOUGH INFORMATION	I DO NOT WANT TO KNOW THIS	HAVE YOU BEEN GIVEN THIS INFORMATION BEFORE?	
					YES	NO
a. Genetics of CF	✓					✓
b. How to treat chest infections		✓			✓	

(EACH ITEM HAS TWO TICKS)

	HOW MUCH INFORMATION WOULD YOU LIKE ABOUT:	A GREAT DEAL MORE INFORMATION	A LITTLE MORE INFORMATION	I HAVE ENOUGH INFORMATION	I DO NOT WANT TO KNOW THIS	HAVE YOU BEEN GIVEN THIS INFORMATION BEFORE?	
						YES	NO
1	How the human body works						
2	Symptoms of CF e.g. coughing						
3	How CF is inherited						
4	Different kinds of treatment for CF						
5	New information, about CF, from other places in the world						
6	Possible complications of CF						
7	How to deal with your feelings about having a brother/sister with CF e.g. sad, angry feelings						
8	What to tell your friends about CF						
9	How to talk to your parents about CF						
10	How serious your brother/sister's illness is						
11	The chances of your children having CF one day						
12	What to expect if your brother/sister's illness gets worse						

Are there any other areas in which you would like more information and advice about CF? If so, please list them in the space below:

____

____

____

____

How accurately do the following statements describe you? Simply indicate how much you agree or disagree with each item, by putting a tick in the column which comes closest to how you feel. Remember that there are no right or wrong answers - I only want your honest opinions. Although some of the statements may seem similar, there are differences between them and each one has been included to help me understand how you feel.

		Strongly Agree	Agree	Neither Agree or Disagree	Disagree	Strongly Disagree
1	I want as much information as possible about CF, good or bad					
2	In uncertain times, I usually hope for the best					
3	I believe in the saying that 'every cloud has a silver lining'					
4	I hardly ever expect good things to happen to me					
5	I always look on the bright side of things					

		Strongly Agree	Agree	Neither Agree or Disagree	Disagree	Strongly Disagree
6	If something can go wrong for me, it will					
7	I hardly ever expect things to go my way					
8	It is my right to know everything about my brother/ sister's illness					
9	I'm always optimistic about the future					
10	Things never work out the way I want them to					
11	Knowing all the facts about CF will only make me miserable					

I am sure that you must think about your CF brother/sister quite a lot:

How would you describe his/her health at present?

Only tick one box

Excellent	☐
Very good	☐
Good	☐
Fair	☐
Poor	☐
Very poor	☐

As your brother/sister with CF gets older, do you expect him/her to

Be cured	☐
Be very much better	☐
Be a little better	☐
Remain the same	☐
Be a little worse	☐
Be a lot worse	☐
Be a great deal worse	☐

I'M SURE THAT YOU ARE WONDERING IF THIS QUESTIONNAIRE WILL EVER COME TO AN END! I KNOW IT IS VERY LONG - BUT YOU ARE ALREADY HALF-WAY THROUGH IT

SECTION 3

FINALLY I'D LIKE TO ASK YOU SOME QUESTIONS ABOUT YOURSELF AND YOUR FAMILY.

Firstly, I'd like to ask you about your family:

We know that families can be very different - especially when it comes to how much family members talk to each other and share problems.

Your answers to the next few questions will help me to understand your family a little better.

How much do you depend on your Mom for information and guidance about CF?

- Completely ☐
- Very much ☐
- Quite a bit ☐
- A little ☐
- Not at all ☐

How much do you depend on your Dad for information and guidance about CF?

- Completely ☐
- Very much ☐
- Quite a bit ☐
- A little ☐
- Not at all ☐

How close do the following statements come to describing how you feel about your family?

For each item, place a tick in the column that comes closest to how you feel.

		MOST OF THE TIME	SOME OF THE TIME	HARDLY EVER
1	I am satisfied that I can turn to my family for help when something is troubling me			
2	I am satisfied with the way our family talks things over and shares problems			
3	I am satisfied that my family accepts and supports my wishes to take on new activities or directions			
4	I am satisfied with the way my family expresses affection and responds to my feelings such as anger, sadness or love.			
5	I am satisfied with the way our family shares time together			

What worries YOU most about CF? Answer in your own words.

____

____

What do you feel is the WORST sickness a person can have? Answer in your own words.

____

You're coming very close to the end of the questionnaire!

NOW I HAVE SOME QUESTIONS ABOUT HOW YOU HAVE BEEN FEELING LATELY

The final questions are concerned with your feelings. For each item, place a tick in the column which comes closest to how you have been feeling in the PAST TWO WEEKS.

		MOST OF THE TIME	SOME OF THE TIME	NEVER
1	I still look forward to things as much as I used to			
2	I sleep very well			
3	I feel like crying			
4	I like to have fun			
5	I feel like running away			
6	I get tummy aches			
7	I have lots of energy			
8	I enjoy my food			
9	I can stick up for myself			
10	I feel my life isn't worth living			
11	I am good at the things I do			
12	I enjoy the things I do			
13	I like talking to my family			
14	I have horrible dreams			
15	I feel very lonely			
16	I am easily cheered up			
17	I feel so sad I can hardly stand it			
18	I feel very bored			

Once more could you please indicate whether or not the following statements describe how you feel. Read each one carefully and put a tick in the YES column if you think a statement is true for you. If it does not apply to you, put your tick in the NO column.

		YES this is true for me	NO this is not true for me
1	I have trouble making up my mind		
2	I get nervous when things don't go right for me		
3	Other people seem to do things more easily than I can		
4	I often have trouble getting my breath		
5	I worry a lot of the time		
6	I am afraid of a lot of things		
7	I get angry easily		
8	I worry about what my parents will say		
9	I feel that other people do not like the way I do things		
10	It's hard for me to get to sleep at night		
11	I worry about what other people think of me		
12	I feel alone even when there are people with me		
13	I often have a frightened feeling like 'butterflies' in my tummy		
14	My feelings get hurt easily		
15	My hands often feel sweaty		
16	I am tired a lot		

		YES this is true for me	NO this is not true for me
17	I worry about what is going to happen		
18	Other children/people are happier than I am		
19	I have bad dreams		
20	My feelings get hurt easily when I am moaned at		
21	I feel that people will tell me I do things the wrong way		
22	I sometimes wake up scared		
23	I worry when I go to bed at night		
24	It is hard for me to keep my mind on my school work/work		
25	I can't sit still for long		
26	I am nervous		
27	A lot of people are against me		
28	I often worry about something awful happening to me		

If you have anything else to add, please use the space below. I would really appreciate your comments and suggestions.

Thank you very much for completing this questionnaire.

APPENDIX 4

LOCATION OF STANDARDIZED SCALES ON THE ORIGINAL QUESTIONNAIRES ACCORDING TO FAMILY GROUPING[1]

	HOSPITAL ANXIETY AND DEPRESSION SCALE	SELF-RATING DEPRESSION SCALE	REVISED CHILDREN'S MANIFEST ANXIETY SCALE	THE LIFE ORIENTATION TEST (DISPOSITIONAL OPTIMISM)	FAMILY APGAR (FAMILY FUNCTIONING)
PARENTS	Pages 24-26[2] 14 items			Pages 20-21 8 items (Nos 1,2,4,6,7,10, 11,14)	Page 23 5 items
PATIENTS		Page 25 18 items	Pages 26-27 28 items	Pages 21-22 8 items (Nos 1,2,4,6,7,10 11,14)	Page 24 5 items
SIBLINGS		Page 20 18 items	Pages 21-22 28 items	Pages 15-16 8 items (Nos 2,3,4,5,6,7, 8,9)	Page 19 5 items

---

[1] A blank space indicates that the scale was not included in the questionnaire
[2] Page numbers refer to those at the bottom of the pages on the 3 sets of questionnaires.

APPENDIX 5

LOCATION OF UNSTANDARDIZED MEASURES ON THE ORIGINAL QUESTIONNAIRES ACCORDING TO FAMILY GROUPING[1]

	CF KNOWLEDGE TEST	PERCEPTION OF PRESENT AND FUTURE HEALTH	INFORMATION PREFERENCES	INFORMATION NEEDS[2]	SOURCES OF INFORMATIONAL SUPPORT	DOCTOR-PARENT/ PATIENT COMMUNICATION PROBLEMS
PARENTS	Pages 3-11[3] 63 items	Page 22 2 items	Page 16 3 items	Pages 14-15 20 items	Pages 17-18 12 items	Pages20-21 8 items (Nos 3,5,8,9,12, 13,15,16)
PATIENTS	Pages 3-11 63 items	Page 17 2 items	Page 16 3 items	Pages 14-15 19 items	Pages 18-19 13 items	Pages 22-23 8 items (Nos 3,5,8,9,12, 13,15,16)
SIBLINGS	Pages 3-11 63 items	Page 17 2 items	Pages 15-16 3 items (Nos 1,8,11)	Page 14 12 items		

---

[1] A blank space indicates that the measure was not included in the questionnaire

[2] Respondents also had to indicate whether they had been given this information before

[3] Page numbers refer to those at the bottom of the pages on the 3 sets of questionnaires.

APPENDIX 6

READABILITY LEVELS OF ENGLISH QUESTIONNAIRES

The readability levels of the English questionnaires were calculated according to the Flesch Formula (Spadaro, Robinson and Smith 1980). The analysis is based on the average sentence length (in words) of selected samples and on the average word length measured as syllables per 100 words of the sample. These two variables are combined to yield a reading ease score (RE).

$$RE = 206.835 - 1.015\ SL - 0.846\ WL$$

where RE = Reading ease score

SL = Average sentence length in words calculated by counting the number of words in the sample and dividing by the number of sentences

WL = Average word length calculated by counting all the syllables in the sample as if reading the words aloud. The syllables are then divided by the number of words in the sample and multiplied by 100 (Spadaro, Robinson and Smith 1980).

The Flesch formula was applied to the following sections of the English questionnaires:

Parents:	Page 1, paragraphs 2 and 3
	Page 12, paragraphs 4, 5 and 6
Patients:	Page 1, paragraphs 1 and 2
	Page 12, all paragraphs
Siblings:	Page 1, paragraphs 1 and 2
	Page 12, all paragraphs

APPENDIX 7

LOCATION OF 63 ITEMS ON THE CF KNOWLEDGE TEST ACCORDING TO SUB-SECTION CONTENT AND COGNITIVE TASKS

	COGNITIVE TASKS[1]		TOTAL	
	RECALL OF KNOWLEDGE	APPLICATION OF KNOWLEDGE	n	%
General CF Knowledge	1,2,3,4,5*,7,10*,14*,27,47,48,55,56		13	20.63
Respiratory Symptoms and Treatment	6,12,13*,15*	26	5	7.94
Physiotherapy	19*,20,21*,22,23,24	25	7	11.11
GIT Symptoms and Treatment	28,29,30,32*,33		5	7.94
Nutrition	34*,35,37,38,41,42,43,45*,46	36,39,40,44	13	20.63
Genetics	49,50,51	52,53,54	6	9.52
Terminology	8,9,31		3	4.76
Anatomy	57,58,59,60,61,62,63		7	11.11
General Medical Knowledge	11,16,17,18		4	6.35

1 Item numbers as they appear in the questionnaires
* Commonly held misconceptions

APPENDIX 8

TEST SCORES (% CORRECT RESPONSES) ACCORDING TO SUB-SECTIONS ON CF KNOWLEDGE TEST FOR PAEDIATRICIANS, PHYSIOTHERAPISTS, NURSING SISTERS, IVTH YEAR MEDICAL STUDENTS, NON-MEDICAL HOSPITAL PERSONNEL AND MOTHERS OF CF PATIENTS

CF KNOWLEDGE TEST	PAEDIATRICIANS AND PAEDIATRIC REGISTRARS	PHYSIOTHERAPISTS	NURSING SISTERS	IVTH YEAR MEDICAL STUDENTS	MOTHERS OF CF PATIENTS	NON-MEDICAL HOSPITAL PERSONNEL
	n = 18	n = 8	n = 8	n = 32	n = 60	n = 14
TOTAL TEST SCORES	97.35	88.10	76.39	82.80	73.10	51.36
SUB-SECTION SCORES General CF Knowledge	91.88	87.50	75.96	84.14	83.97	58.24
Respiratory Symptoms and Treatment	100.00	90.00	67.50	79.38	74.33	34.29
Physiotherapy	97.62	96.43	82.14	77.68	90.48	57.14
GIT Symptoms & Treatment	92.22	80.00	62.50	63.13	81.67	21.43
Nutrition	97.86	83.65	74.04	81.01	71.41	56.59
Genetics	98.15	70.83	50.00	70.84	66.67	21.43
Anatomy	100.00	100.00	100.00	100.00	85.48	83.67
CF Terminology	100.00	100.00	100.00	100.00	46.11	33.33
General Medical Knowledge	97.22	96.88	78.13	89.06	60.83	55.36

APPENDIX 9
SUMMARY DATA ON CF KNOWLEDGE TEST FOR MARRIED MOTHERS[1]

	No OF ITEMS	MEAN[2]		SD	RANGE
Composite Test Score	63	48.04	(76.25)	8.73	24 - 62
Revised Test Score	56	44.04	(78.64)	7.38	24 - 55
Sub-Section Scores					
General CF knowledge	13	11.28	(86.77)	1.87	2 - 13
Respiratory symptoms and treatment	5	3.85	(77.04)	1.11	1 - 5
Physiotherapy	7	6.43	(91.86)	1.02	1 - 7
GIT symptoms and treatment	5	4.28	(85.60)	1.38	0 - 5
Nutrition	13	9.72	(74.77)	2.93	0 - 13
Genetics	6	4.22	(70.33)	1.95	0 - 6
Anatomy	7	5.98	(85.43)	1.97	0 - 7
Terminology	3	1.46	(48.67)	1.21	0 - 3
General Medical	4	2.54	(63.50)	1.17	0 - 4

SUB-SECTION SCORES ACCORDING TO INDIVIDUAL ITEMS[3]

CF GENERAL		RESP SYMP & TREATMENT		PHYSIOTHERAPY		GIT SYMP & TREATMENT		NUTRITION		GENETICS		ANATOMY		TERMINOLOGY		GENERAL MEDICAL	
1[4]	100.00	6	98.15	19	90.50	28	90.74	34	55.56	49	88.89	57	96.30	8	37.04	11	57.41
2	92.59	12	87.04	20	85.19	29	88.89	35	72.22	50	72.22	58	88.89	9	51.85	16	87.04
3	100.00	13	77.78	21	90.74	30	87.04	36	53.70	51	70.37	59	79.63	31	57.41	17	46.30
4	87.04	15	90.74	22	94.44	32	85.19	37	70.37	52	81.48	60	81.48			18	62.96
5	98.15	26	31.48	23	98.15	33	75.93	38	96.30	53	44.44	61	79.63				
7	90.74			24	96.30			39	57.41	54	66.67	62	87.04				
10	59.96			25	87.03			40	94.44			63	85.19				
14	24.07							41	83.33								
27	94.44							42	88.89								
47	92.59							43	77.78								
48	98.15							44	79.63								
55	98.15							45	48.15								
56	92.59							46	94.44								

---

[1] n = 54

[2] Raw Scores with percentages in brackets

[3] % Correct Responses only

[4] Item numbers as they appear on the questionnaire

APPENDIX 10

MEANS, SD's AND RANGES (RAW SCORES) ON 9 SUB-SECTIONS OF THE CF KNOWLEDGE TEST ACCORDING TO FAMILY GROUPING[1]

	No OF ITEMS[2]	MEAN	SD	RANGE
GENERAL CF KNOWLEDGE	13			
Fathers		10.91	1.39	8 - 13
Mothers		10.92	1.87	2 - 13
Patients		10.44	1.72	6 - 13
Siblings		9.59	1.94	3 - 12
RESPIRATORY SYMPTOMS & TREATMENT	5			
Fathers		3.48	1.09	1 - 5
Mothers		3.72	1.11	1 - 5
Patients		3.39	0.78	2 - 4
Siblings		3.03	1.12	1 - 5
PHYSIOTHERAPY	7			
Fathers		6.07	1.01	1 - 7
Mothers		6.33	1.16	1 - 7
Patients		5.83	1.34	3 - 7
Siblings		5.07	1.49	1 - 7
GIT SYMPTOMS AND TREATMENT	5			
Fathers		4.17	1.00	1 - 5
Mothers		4.08	1.38	0 - 5
Patients		4.50	0.62	3 - 5
Siblings		3.66	1.39	0 - 5
NUTRITION	13			
Fathers		9.33	2.67	4 - 13
Mothers		9.28	2.93	0 - 13
Patients		9.06	2.44	4 - 13
Siblings		8.07	2.69	1 - 13
GENETICS	6			
Fathers		4.22	1.95	0 - 6
Mothers		4.00	1.95	0 - 6
Patients		2.83	1.82	0 - 6
Siblings		2.69	1.87	0 - 6
ANATOMY	7			
Fathers		5.89	1.69	0 - 7
Mothers		5.98	1.97	0 - 7
Patients		6.28	1.23	3 - 7
Siblings		5.62	2.11	0 - 7
TERMINOLOGY	3			
Fathers		1.30	1.41	0 - 3
Mothers		1.38	1.21	0 - 3
Patients		1.28	1.18	0 - 3
Siblings		0.90	0.90	0 - 3
GENERAL MEDICAL KNOWLEDGE	4			
Fathers		2.24	1.23	0 - 4
Mothers		2.43	1.67	0 - 4
Patients		2.05	1.61	0 - 4
Siblings		1.83	1.10	0 - 4

---

[1] n = 54 fathers n = 60 mothers n = 18 patients n = 29 siblings

[2] Number of items in each sub-section

APPENDIX 11

COMPOSITE CF KNOWLEDGE OF FATHERS AND MOTHERS VERSUS SELECTED PATIENT VARIABLES[1]

	p VALUES	
	FATHERS	MOTHERS
Age	0.2149	0.3258
Shwachman score	0.4742	0.3582
Period since diagnosis	0.5934	0.0456
Days in hospital	0.4705	0.2670
No of clinic attendances[2]	-	0.1034

1 n = 54 fathers n = 60 mothers
2 Not calculated for fathers

APPENDIX 12

COMPOSITE CF KNOWLEDGE OF PATIENTS AND SIBLINGS VERSUS SELECTED SOCIODEMOGRAPHIC AND MEDICAL VARIABLES

	p VALUES			
	PATIENTS[1]		SIBLINGS[2]	
	MALE	FEMALE	MALE	FEMALE
Education	0.1244	0.4200	0.4847	0.5858
Shwachman score	0.2489	0.1070	0.5392	0.9452
Period since diagnosis	0.1427	0.7302	0.8787	0.4147
Days in hospital[3]	0.0082*	0.3628		
Number of clinic attendances[3]	0.5201	0.6418		

1 n = 9 males 9 females
2 n = 13 males 16 females
3 Not calculated for siblings

APPENDIX 13

PERCEPTION OF THE CF PATIENT'S PRESENT AND FUTURE HEALTH VERSUS SELECTED SOCIODEMOGRAPHIC AND MEDICAL VARIABLES ACCORDING TO FAMILY GROUPING

	p VALUES					
	PRESENT HEALTH[1]			FUTURE HEALTH[2]		
	PARENTS	PATIENTS	SIBLINGS	PARENTS	PATIENTS	SIBLINGS
Age	0.4466	0.5854	0.4181	0.4373	0.4660	0.5814
Education		0.5486	0.4173		0.3348	0.3311
Shwachman score	0.9232	0.3197	0.1747	0.6129	0.1314	0.1692
Period since diagnosis	0.5727	0.3745	0.7887	0.1038	0.1000	0.5681
Days in hospital	0.0959	0.5763		0.5990	0.1912	
Number of clinic attendances[3]	0.4052	0.1386		0.5284	0.1345	

1 n = 114 parents n = 18 patients n = 29 siblings
2 n = 110 parents n = 18 patients n = 29 siblings
3 n = 60 mothers

APPENDIX 14

PAIRED ANALYSIS OF PATIENT AND SIBLING PERFORMANCE ON SELECTED ITEMS FROM THE REVISED CHILDREN'S MANIFEST ANXIETY SCALE AND THE DEPRESSION SELF RATING SCALE[1]

ITEM	CHI-SQUARE	p VALUE
ANXIETY (RCMAS)		
I often have trouble getting my breath	$\text{Chi}^2 = 2.67$	0.1025
It's hard for me to get to sleep at night	$\text{Chi}^2 = 1.00$	0.3173
My hands often feel sweaty at night	$\text{Chi}^2 = 4.50$	0.0339
I am tired a lot	$\text{Chi}^2 = 0.00$	1.0000
DEPRESSION (DSRS)[2]		
I sleep very well	$\text{Chi}^2 = 0.33$	0.5637
I get tummy aches	$\text{Chi}^2 = 0.00$	0.1000
I have lots of energy	$\text{Chi}^2 = 0.20$	0.6547
I enjoy my food	$\text{Chi}^2 = 0.20$	0.6547

---

[1] n = 13 pairs

[2] Response options were dichotomized as follows: most of the time, some of the time/never.

APPENDIX 15

INFORMATION PREFERENCES VERSUS SELECTED SOCIODEMOGRAPHIC AND MEDICAL VARIABLES ACCORDING TO FAMILY GROUPING[1]

	p VALUES			
	FATHERS	MOTHERS	PATIENTS	SIBLINGS
Age	0.9787	0.5010	0.2531	0.4911
Social class	0.4194	0.1938	0.2071	0.8093
Schwachman score	0.5638	0.1695	0.6981	0.4400
Days in hospital[2]	0.5531	0.9251	0.7887	
Number of clinic attendances[3]		0.7477	0.2021	

1 n = 54 fathers n = 60 mothers n = 18 patients n = 28 siblings
2 Not calculated for siblings
3 Not calculated for fathers and siblings

APPENDIX 16

MEANS, SD's AND RANGES OF SCORES ON THE INDEX MEASURE OF INFORMATION NEEDS ACCORDING TO FAMILY GROUPING[1]

	n	MEAN	SD	POSSIBLE RANGE	MID-POINT	OBTAINED RANGE
Fathers	53	44.68	10.20	20 - 60	40	20 - 60
Mothers	60	42.40	10.50	20 - 60	40	20 - 60
Patients	18	31.94	10.82	19 - 57	38	19 - 52
Siblings	28	24.29	8.97	12 - 36	24	12 - 36

1 Information needs were measured on a 3 point scale (3 = a great deal more information, 2 = a little more information and 0 = I have enough information)

APPENDIX 17

MEANS, SD's AND RANGES OF SCORES ON THE INDEX MEASURE OF INFORMATION NOT GIVEN BEFORE ACCORDING TO FAMILY GROUPING[1]

	n	MEAN	SD	POSSIBLE RANGE	MID-POINT	OBTAINED RANGE
Fathers	53	29.19	5.88	20 - 40	30	20 - 40
Mothers	60	28.75	5.03	20 - 40	30	20 - 40
Patients	18	27.22	5.37	19 - 38	28.5	19 - 37
Siblings	28	18.64	3.64	12 - 24	18	12 - 24

[1] Information Not Given Before was assessed by means of a dichotomous Yes or No response (Yes = 1, No = 2)

APPENDIX 18

PERCENTAGE DISTRIBUTION OF FATHERS' NEEDS FOR FURTHER INFORMATION ON SELECTED MEDICAL AND PSYCHOSOCIAL TOPICS[1]

	TOPICS	A GREAT DEAL MORE INFORMATION	A LITTLE MORE INFORMATION	I HAVE ENOUGH INFORMATION
1	How the human body works	30.19	37.74	32.07
2	Symptoms of CF	43.40	20.75	35.85
3	How CF is inherited	28.30	22.64	49.06
4	When and how to administer antibiotics	39.62	41.51	18.87
5	When and how to take pancreatic enzymes	24.53	39.62	35.85
6	How to perform physiotherapy	26.42	43.40	30.18
7	How to use a nebulizer	35.85	37.74	26.41
8	Forms of exercise and sport that are suitable for people with CF	50.94	24.53	24.53
9	Healthy meals for CF children	49.06	33.96	16.98
10	New information, about CF, from other places in the world	79.25	15.09	5.65
11	Detailed results of any tests performed on your child when he/she attends clinic e.g. X rays, sputum tests	52.83	28.30	18.87

Continued on next page

Appendix 18 continued

	TOPICS	A GREAT DEAL MORE INFORMATION	A LITTLE MORE INFORMATION	I HAVE ENOUGH INFORMATION
12	Possible complications of CF	67.92	20.75	11.32
13	How to deal with your child's feelings about having CF e.g. feelings of anger, sadness, frustration	56.60	24.53	18.87
14	What to tell your child about his/her illness	43.40	35.85	20.75
15	What to tell your other children about CF	47.17	13.21	39.63
16	How CF might affect your child's career, social life and marriage	67.92	13.21	18.87
17	How CF might affect your child's chances of having his/her own children	64.15	18.87	16.98
18	How serious your child's illness is	58.49	15.09	26.42
19	What to expect if your child's illness gets worse	60.38	24.53	15.09
20	How to answer your child's questions about whether he might die from his illness	50.94	22.64	26.42

1 n = 53

APPENDIX 19

PERCENTAGE DISTRIBUTION OF MOTHERS' NEEDS FOR FURTHER INFORMATION ON SELECTED MEDICAL AND PSYCHOSOCIAL TOPICS[1]

	TOPICS	A GREAT DEAL MORE INFORMATION	A LITTLE MORE INFORMATION	I HAVE ENOUGH INFORMATION
1	How the human body works	33.33	30.00	36.67
2	Symptoms of CF	26.67	25.00	48.33
3	How CF is inherited	20.00	23.33	56.67
4	When and how to administer antibiotics	36.67	25.00	38.33
5	When and how to take pancreatic enzymes	10.00	26.67	63.33
6	How to perform physiotherapy	16.67	31.67	51.67
7	How to use a nebulizer	38.33	23.33	38.33
8	Forms of exercise and sport that are suitable for people with CF	33.33	41.67	25.00
9	Healthy meals for CF children	35.00	36.67	28.33
10	New information, about CF, from other places in the world	68.33	28.33	3.33
11	Detailed results of any tests performed on your child when he/she attends clinic e.g. X rays, sputum tests	36.67	38.33	25.00

Continued on next page

Appendix 19 continued

	TOPICS	A GREAT DEAL MORE INFORMATION	A LITTLE MORE INFORMATION	I HAVE ENOUGH INFORMATION
12	Possible complications of CF	56.67	21.67	21.67
13	How to deal with your child's feelings about having CF e.g. feelings of anger, sadness, frustration	63.33	30.00	6.67
14	What to tell your child about his/her illness	43.33	38.33	18.34
15	What to tell your other children about CF	40.00	38.33	21.67
16	How CF might affect your child's career, social life and marriage	60.00	26.67	13.33
17	How CF might affect your child's chances of having his/her own children	63.33	20.00	16.67
18	How serious your child's illness is	43.33	31.67	25.00
19	What to expect if your child's illness gets worse	51.67	28.33	20.00
20	How to answer your child's questions about whether he might die from his illness	61.67	23.33	15.00

1 n = 60

APPPENDIX 20

INDEX MEASURES OF INFORMATION NEEDS AND INFORMATION NOT GIVEN BEFORE VERSUS SELECTED SOCIODEMOGRAPHIC AND MEDICAL VARIABLES ACCORDING TO FAMILY GROUPING[1]

	p Values					
	INFORMATION NEEDS			INFORMATION NOT GIVEN BEFORE		
	PARENTS	PATIENTS	SIBLINGS	PARENTS	PATIENTS	SIBLINGS
Age	0.1029	0.2046	0.8912	0.1652	0.0235	0.4244
Education[2]		0.1206	0.8094		0.1302	0.7632
Social class	0.0009*	0.1645	0.1743	0.0001*	0.1012	0.2034
Shwachman score	0.7531	0.0414	0.4701	0.6077	0.3095	0.8905
Period since diagnosis	0.0618	0.2167	0.6077	0.3738	0.4203	0.5413
Days in hospital[3]	0.9875	0.1032		0.7803	0.2513	
Number of clinic attendances[4]	0.9783	0.1274		0.3154	0.3163	

1 n = 113 parents n = 18 patients n = 28 siblings
2 Not calculated for parents
3 Not calculated for siblings
4 Not calculated for fathers and siblings

APPENDIX 21

PARENTAL RATINGS OF FREQUENCY OF USE (% DISTRIBUTION) OF SOURCES OF INFORMATIONAL SUPPORT AND GUIDANCE ABOUT CF[1]

		MANY TIMES	OCCASIONALLY	NOT AT ALL	FAMILY GROUPING
1	Other parents who also have a child with CF	7.41	53.70	38.89	Fathers
		20.00	58.33	21.67	Mothers
2	Your family doctor	9.26	24.07	66.67	Fathers
		13.33	26.67	60.00	Mothers
3	The doctors at the CF clinic	44.44	35.19	20.37	Fathers
		70.00	28.33	1.67	Mothers
4	The physiotherapists at the hospital	12.96	31.48	55.56	Fathers
		11.67	36.67	51.67	Mothers
5	The social worker at the CF clinic	1.85	12.96	85.19	Fathers
		5.00	31.67	63.33	Mothers
6	The dietician at the hospital	3.70	14.81	81.48	Fathers
		3.33	8.33	88.33	Mothers
7	Books, pamphlets and magazines about CF	35.18	35.19	29.63	Fathers
		43.33	40.00	16.67	Mothers
8	The CF Association	12.96	46.30	40.74	Fathers
		26.67	28.33	45.00	Mothers
9	Prayer and Faith	55.56	18.52	25.92	Fathers
		66.67	18.33	15.00	Mothers

Continued on next page

Appendix 21 continued

		MANY TIMES	OCCASIONALLY	NOT AT ALL	
10	Your priest or minister	24.07	14.81	61.11	Fathers
		21.67	25.00	53.33	Mothers
11	Your husband or wife	66.67	31.48	1.85	Fathers
		40.00	35.00	25.00	Mothers
12	Your local pharmacy	3.70	14.81	81.48	Fathers
		8.33	11.67	80.00	Mothers

1 n = 54 fathers n = 60 mothers

APPENDIX 22

PARENTAL RATINGS OF PERCEIVED HELPFULNESS (% DISTRIBUTION) OF SOURCES OF INFORMATIONAL SUPPORT AND GUIDANCE ABOUT CF[1]

		A GREAT DEAL	QUITE A LOT	NOT VERY MUCH	NONE AT ALL	FAMILY GROUPING
1	Other parents who also have a child with CF	7.41	27.78	33.33	31.48	Fathers
		20.00	26.67	26.67	26.69	Mothers
2	Your family doctor	3.70	12.96	24.07	59.26	Fathers
		1.67	16.67	21.67	60.00	Mothers
3	The doctors at the CF clinic	40.74	35.18	7.41	16.67	Fathers
		45.00	41.67	13.33	-	Mothers
4	The physiotherapists at the hospital	11.11	25.92	7.41	55.56	Fathers
		11.67	18.33	30.00	40.00	Mothers
5	The social worker at the CF clinic	1.85	11.11	12.96	74.07	Fathers
		-	15.00	25.00	60.00	Mothers
6	The dietician at the hospital	1.85	11.11	11.11	75.93	Fathers
		-	3.33	8.33	88.83	Mothers
7	Books, pamphlets and magazines about CF	25.92	31.48	14.81	27.78	Fathers
		28.33	31.67	26.67	13.33	Mothers
8	The CF Association	9.26	38.89	14.81	37.04	Fathers
		13.33	33.33	11.67	41.67	Mothers

Continued on next page

Appendix 22 continued

		A GREAT DEAL	QUITE A LOT	NOT VERY MUCH	NONE AT ALL	FAMILY GROUPING
9	Prayer and Faith	44.44	22.22	9.26	24.07	Fathers
		53.33	18.33	13.33	15.00	Mothers
10	Your priest or minister	14.81	9.26	11.11	64.81	Fathers
		18.33	11.67	15.00	55.00	Mothers
11	Your husband or wife	46.30	40.74	11.11	1.85	Fathers
		33.33	26.67	15.00	25.00	Mothers
12	Your local pharmacy	-	3.70	12.96	83.33	Fathers
		1.67	5.00	15.00	78.33	Mothers

1 n = 54 fathers n = 60 mothers

APPENDIX 23

PATIENTS' RATINGS OF FREQUENCY OF USE (% DISTRIBUTION) OF SOURCES OF INFORMATIONAL SUPPORT AND GUIDANCE ABOUT CF[1]

		MANY TIMES	OCCASIONALLY	NOT AL ALL
1	Other people who also have CF	16.67	16.67	66.67
2	Your family doctor	5.56	16.67	77.77
3	The doctors at the CF clinic	83.33	11.11	5.56
4	The physiotherapists at the hospital	5.56	44.44	50.00
5	The social worker at the CF clinic	11.11	16.67	72.22
6	The dietician at the hospital	-	16.67	83.33
7	Books, pamphlets and magazines about CF	22.22	38.89	38.89
8	The CF Association	5.56	22.22	72.22
9	Prayer and Faith	61.11	22.22	16.67
10	Your priest or minister	22.22	11.11	66.67
11	Your Mom	72.22	22.22	5.56
12	Your Dad	38.89	27.78	33.33
13	Your Chemist	-	5.56	94.44

[1] n = 18 patients

APPENDIX 24

PATIENTS' RATINGS OF PERCEIVED HELPFULNESS (% DISTRIBUTION) OF INFORMATIONAL SUPPORT AND GUIDANCE ABOUT CF[1]

		A GREAT DEAL	QUITE A LOT	NOT VERY MUCH	NONE AT ALL
1	Other people who also have CF	-	5.56	33.33	61.11
2	Your family doctor	5.56	5.56	22.22	66.67
3	The doctors at the CF clinic	33.33	55.55	5.56	5.56
4	The physiotherapist at the hospital	-	11.11	27.78	61.11
5	The social worker at the CF clinic	5.56	-	16.67	77.78
6	The dietician at the hospital	-	5.56	11.11	83.33
7	Books, pamphlets and magazines about CF	22.22	16.67	27.78	33.33
8	The CF Association	5.56	11.11	22.22	61.11
9	Prayer and Faith	55.55	16.67	11.11	16.67
10	Your priest or minister	16.67	5.56	16.67	61.11
11	Your Mom	66.67	22.22	5.56	5.56
12	Your Dad	38.89	16.67	11.11	33.33
13	Your Chemist	-	-	5.56	94.44

[1] n = 18 patients

APPENDIX 25

INDEX OF RATINGS OF FREQUENCY OF USE AND DEGREE OF HELPFULNESS OF SOURCES OF INFORMATIONAL SUPPORT AND GUIDANCE ABOUT CF VERSUS SELECTED SOCIODEMOGRAPHIC AND MEDICAL VARIABLES ACCORDING TO FAMILY GROUPING[1]

	p VALUES					
	FREQUENCY OF USE			DEGREE OF HELPFULNESS		
	FATHERS	MOTHERS	PATIENTS	FATHERS	MOTHERS	PATIENTS
Age	0.8793	0.7220	0.9296	0.7684	0.8188	0.6579
Social class	0.6013	0.8424	0.1098	0.8559	0.4064	0.9983
Shwachman score	0.5378	0.1855	0.3260	0.6759	0.4680	0.0696
Period since diagnosis	0.7774	0.7447	0.8299	0.7165	0.8698	0.8396
Days in hospital	0.5727	0.2816	0.6059	0.9737	0.1471	0.2655
Number of clinic attendances[2]		0.6800	0.5246		0.4961	0.1089

[1] n = 54 fathers n = 60 mothers n = 18 patients
[2] Not calculated for fathers

APPENDIX 26

INDEX RATINGS OF COMMUNICATION PROBLEMS VERSUS SELECTED SOCIODEMOGRAPHIC AND MEDICAL VARIABLES ACCORDING TO FAMILY GROUPING[1]

	p VALUES	
	PARENTS	PATIENTS
Age	0.2813	0.0409
Language	0.5073	0.2203
Social class	0.0042*	0.1238
Education[2]		0.0208
Shwachman score	0.6404	0.2165
Period since diagnosis	0.8717	0.4004
Days in hospital	0.3031	0.4106
Number of clinic attendances[3]	0.7771	0.9820

[1] n = 114 parents n = 18 patients

[2] Not calculated for parents

[3] Not calculated for fathers

APPENDIX 27

SUMMARY DATA OF SUB-SCALE SCORES ON THE REVISED CHILDREN'S MANIFEST ANXIETY SCALE FOR PATIENTS AND SIBLINGS

SUB-SCALES	MEAN	SD	POSSIBLE RANGE	OBSERVED RANGE
Physiological[1]				
Patients	3.89	2.78	0 - 10	0 - 9
Siblings	3.24	2.41	0 - 10	0 - 8
Worry/Oversensitivity[2]				
Patients	3.89	3.38	0 - 11	0 - 11
Siblings	4.90	3.61	0 - 11	0 - 11
Concentration[3]				
Patients	1.94	1.62	0 - 7	0 - 5
Siblings	1.97	2.15	0 - 7	0 - 6

[1] Item Numbers: 1, 4, 7, 10, 13, 15, 16, 19, 22, 25

[2] Item Numbers: 2, 5, 6, 8, 11, 14, 17, 20, 23, 26, 28

[3] Item Numbers: 3, 9, 12, 18, 21, 24, 27

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