GETTING ON WITH LIFE: THE LIVED EXPERIENCE OF FOUR ADULTS WITH CYSTIC FIBROSIS

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ABSTRACT

Cystic fibrosis (CF) is a progressive, life limiting disease that requires relentless life long management. Although there are a small number of adults with CF in New Zealand, the disease is significant because of the duration and intensity of the health care they require.

A phenomenological hermeneutic approach has been used to explore the lived experience of a small group of adults with CF. Their experience is primarily one of getting on with life rather than dwelling on illness. Knowing that they have the disease all their lives leads to a difference in experience from people who are diagnosed with a chronic illness in adulthood. They experience a gradual realising of CF when they begin to understand the meaning of the disease, and after they have experienced significant disease related changes in their bodies. Adolescence and young adulthood are times of learning to live with CF. They learn their own limits, learn to look after themselves and manage their illness. The desire to get on with life leads people with CF to minimise illness to themselves and to others, and to reject aspects of the patient role. They see themselves as being relatively healthy much of the time. This is a different way of looking at health and illness from the traditional disease focused approach.

Nurses and other health professionals can use this new understanding of living with CF to better support these people to lead meaningful and purposeful lives rather than to simply look after their disease. As a consequence of this study, an increasing move to home care with an enhanced continuity of care across of hospital and community settings by health professionals who understand the illness experience are advocated.

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CONTENTS

INTRODUCTION

This study uses a phenomenological hermeneutic approach to uncover and reflect upon the experience of a small group of adults who are living with cystic fibrosis (CF). The aim of phenomenology is to gain a deeper understanding of the nature and meaning of our everyday experiences. In practice this approach meant gaining knowledge of the experience from people with CF, and using their accounts to describe and understand the way they live in the world as an adult with this disease.

Nursing is about caring for people when they are sick but also about helping them in their response to illness. The small number of adults with CF in New Zealand means that most nurses and other health professionals have a limited understanding of the disease itself, let alone the patient's illness experience. The aim of this study is to gain a greater understanding of how adults live with CF, how they create meanings and cope with their disease. It is hoped that as a result of this study, health professionals will gain a greater awareness of this illness experience and will incorporate this understanding into their practice.

CF is a genetic condition. Therefore adults with CF have been living with a chronic disease from birth. The disease is progressive with a reduced life expectancy. Until relatively recently, it was rare for people with the disease to survive to adulthood. In addition to its effects on the body, an ongoing experience of this kind can have a profound effect on the person and family, both economically and emotionally, and on the way they conduct their everyday lives.

Management of the disease is unrelenting and lifelong. It includes frequent chest physiotherapy, daily dietary supplements, pancreatic enzymes and vitamins, antibiotics as required, and regular medical check ups. Despite improvements in therapy and diet, CF remains a significant health problem,

and most adults with the disease are "high users" of health care services. Although there are only a small number of adults with CF in New Zealand, the disease is significant because of the intensity and duration of health care they require. Information collected on the New Zealand National Cystic Fibrosis (CF) database reveals that 94 people out of a total of 302 with the disease are older than 15 years (Wesley, Dawson, Hewitt & Kerr, 1993). Because of the small number of people with CF, viable specialised clinics have not been established on a regional basis in this country. Wesley and Stewart (1985), in their discussion of New Zealand's poorer survival rate compared with some overseas centres, project that in New Zealand there is a 47% probability of survival to 20 years compared to 80% in Victoria, Australia, 67% for Canadian males and 62% in England, Wales and Denmark. Also, they comment on the variable level of expertise available in the settings where people with CF currently receive their medical care.

The experience of living with CF would be expected to have some aspects in common with other chronic illness experiences, but also to have distinctive features specific to this disease. Like other chronic illnesses, management of CF mostly takes place in the patient's home, and it is only during acute exacerbations or progressive deterioration that the person is hospitalised. Adults, who have lived through childhood and adolescence with a life-threatening disease, must learn to cope with the interrelated demands of their illness and their social circumstances, and to create a meaningful life for themselves.

As a result of nursing adults with CF, I became aware that there are distinctive features to their illness experience and the strategies that they use to cope with their illness. In many ways they did not appear to be like other patients. In particular, their medical treatment was distinctive because of the special diet they required and the type and length of time they received intravenous antibiotic therapy. What was even more

noticeable was their approach to hospitalisation; they did not want to lie in bed and passively receive their treatments; they wanted to continue their normal activities and, at times, this created conflict with health professionals. I also became aware that the care provided in acute wards did not seem to meet many of the needs of people with a lifelong respiratory disease. It was not considered possible to provide some medical treatments at home, and there was little understanding or support for the psychosocial impact of the disease. It seemed to me that some readmissions to the ward could be prevented, or the length of stay could be reduced, by providing specialist respiratory nursing in the community.

As part of a small study[1] of how people live with chronic illness, I interviewed some adults with CF. They had a strong desire to be treated normally; they had lived with this disease all their lives and did not want to be treated as sick people. Hospitalisation was required for intravenous antibiotic administration but this did not mean they considered themselves to be sick. Issues about the progressive nature of the disease were touched on but not elaborated. I wondered whether this strong urge not to let the disease make them into sick people underlay their coping and provided the impetus for their prolonged survival. It also appeared that some improvement could be made in the health care received by both the person and their family.

From the outset, some aspects of living with CF discussed by some of the fieldwork participants were expected to be significant in this study: fulfilling an adult role in society, coping with infertility, bringing up children, relationship to spouse and socio-economic aspects of living with a chronic illness. However, this study has revealed other aspects of living day by day with a chronic and progressive illness and does not focus strongly on the above topics. It is important to note that this study does not encompass the

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[1] A fieldwork, exploring the experience of chronic illness, completed as an assignment for a Masters' paper in 1993.

very progressive phase of the disease. While this study does not attempt to define the lived experience of all adults with CF, it has uncovered some patterns in the responses of four adults with CF, and suggests strategies nurses and other health professionals should consider when working with this group of people.

This introduction has provided an overview of the study. Chapter One will review selected literature about CF and the experience of chronic illness. Chapter Two will explain the research method and the way this research was conducted, and includes information about the participants, the ethical considerations, and the method used to obtain and analyse the data. Chapters Three to Six will describe the research findings with themes and associated subthemes in each chapter. Chapter Seven contains a discussion of the key findings in the light of current literature and addresses the implications and limitations of the study.

CHAPTER ONE
LITERATURE REVIEW

Improvements in the management and treatment of many diseases has resulted in an increasing number of people living with a chronic illness. Understanding the experience and effects of chronic illness on the person has been increasing over the last forty years, particularly because of the growth of naturalistic inquiry. The experience of living with CF can be expected to share some of the same characteristics, concerns and meanings as living with other chronic diseases. This chapter will begin with an examination of selected literature concerning the main approaches to understanding the experience of living with a chronic illness. CF will then be considered as a specific disease process; its incidence, symptoms and current management will be outlined, followed by a discussion of what is known from the literature about the psychosocial impact of CF on adults.

CHRONIC ILLNESS

There are three main approaches to understanding the experience of living with chronic illness in the literature. The sociological concept of illness trajectory; psychological concepts of stress, adaptation and coping; and a growing literature in a number of fields that reflects on the impact of illness on personhood.

Illness Trajectory

In the 1960s, two sociologists, Glaser and Strauss (1965,1968) began studying the experience of dying patients in hospitals. They discovered that most of these patients were chronically ill and began to interview them at home, asking them about their experiences. They were among the first researchers to use insider perspectives in the health context, and to include the subjective experience of individuals involved in situations as part of their research (Conrad, 1990). Strauss and Glaser (1975) were also among

the first to study illness outside institutions. Their research showed that most illness management is undertaken by the person and family in their own home, rather than by health professionals in an institutional setting. They were able to identify a number of key problems that the chronically ill and their families must develop strategies to deal with. These include the prevention and management of medical crises, symptom control, maintenance of treatment regimes, social isolation, adjustment to changes in the course of the disease, attempts to normalise life, and economic effects that require extensive changes in lifestyle as well as family and social relationships. By contrast, Dimond and Jones (1983) suggest that, in an acute illness experience, the doctor primarily manages the episode and the patient tends to be a relatively passive recipient of treatment or cure.

Parsons (1951) defined four features of the sick role in Western society: the individual is not responsible for the condition, is exempt from normal social role obligations, is obligated to get well and is obligated to seek and accept professional care. Aspects of this conceptualised role fit the short term illness experience but are not appropriate when a person has a chronic illness. The chronically ill person does not tend to remain exempt from social obligations for the duration of the illness and may manage many aspects of his or her own care (Dimond & Jones, 1983). Parsons' definition also excludes any appreciation of the permanent physical, social and psychological adjustments the chronically ill person must make.

Strauss and Glaser's (1975) research generated an understanding that the major issues in managing a long-term chronic illness are more likely to be psychosocial than medical, and that the psychosocial effects of an illness can be independent of the course of the disease. Conrad (1990) clarifies the distinction between disease and illness.

> It is by now becoming common in sociology to distinguish between disease and illness. We can see disease as an

> undesirable physiological process or state. Illness by contrast, can be depicted as the social and psychological phenomena that accompany these putative physiological problems. Illness is a profoundly social phenomenon that may or may not rest on disease as a foundation. It has more to do with perception, behaviour and experience than with physiological process (p. 1259).

Dimond and Jones (1983) propose three perspectives on chronic illness. The first considers the clinical or physiological manifestations - how the disease affects the body and the treatment implications; the second is the meaning the illness has to an individual, including the influence of age, sex, cultural and social expectations, and the effects of the illness on lifestyle and role; the third is the definitions of illness that are held by the community, family and acquaintances. These three perspectives interact to determine the nature of the "disability" of a person with chronic illness.

Strauss (1987) argues that the health care system has not kept pace with the needs of the chronically ill because it is still designed primarily for the treatment of acute disease. He believes that a model needs to be developed that "casts the ill and their intimates, if they have them, as the central agents of management and the home as the vital stage where most of the management takes place" (p. 33). According to Strauss, illnesses have different phases, acute management may take place in the hospital, but most illness management actually takes place at home, by the ill person and their family. The ill need more support at home, and health care services should move towards home care.

Strauss and Glaser (1975) developed the conceptual framework of the illness trajectory which is a number of phases within an illness experience including the associated "work" of managing the illness that is undertaken by the person and family. Lubkin (1990) suggests that the concept of illness

trajectory has two foci; one is on the difference in perception, over time, of those involved in the situation and the second is its impact on these people and their responses. This second perspective takes into account the identification, organisation, and performance of tasks involved in management during the entire course of the illness.

Different diseases have different trajectory patterns because of their clinical features and specific management regime. There are also differences between people with the same disease as a result of the progress of the disease, the person's response to it and associated biographical contingencies (Corbin & Strauss, 1988). Biography has three elements: biographical time, conceptions of self, and body. Illness may effect a person's biography by interrupting and altering our story of ourselves and our future, by altering our self image and by affecting our body's ability to carry out the tasks that make up our normal life. "Biographical work" is considered an important part of managing illness. People who are ill must come to terms with "actual or potential failed performances", adjusting their identities and "biographical direction" (Corbin & Strauss, 1988, p. 68).

Corbin and Strauss (1988) identified the phases that can be expected to occur in any chronic illness. These phases may vary in length and they require different "work" depending on the disease and the individual. There is a phase of initial disruption caused by the illness and the process of diagnosis. The work in this phase is associated with uncovering the source of symptoms and waiting for the diagnosis. This is followed by a phase of accommodating the illness: contextualising (incorporating the illness trajectory into biography), coming to terms, reconstituting identity and recasting biography (giving new directions to biography). Stable phases may follow where the management of the disease is incorporated into everyday life. This work not only affects the ill person but also requires the involvement of his or her partner and family. Unstable and downward

phases demand endurance during frequent physical and emotional ups and downs. The person's life is physically, socially and biographically disrupted.

Corbin and Strauss (1991) promote illness trajectory as a framework for nurses to use to understand the experience of chronic illness, and to guide nursing practice and nursing research in this area. Strauss and Glaser (1975) believed that the chronically ill should have greater participation in health care, and that the health care system should be extended to deal with problems of chronicity. Health professionals should spend more time understanding the person rather than doing tasks, become more accountable for psychological and social aspects of care rather than focusing on carrying out medical or medically supportive activities, and take time to understand each patient's biography.

Health professionals' attitudes towards compliance with treatment are slowly changing as the ill person's perspective is better understood. The usual definition of compliance is the extent to which behaviour coincides with medical advice. A study of women's attitudes towards complying with treatments showed that it was not a lack of understanding or inconsistency in explanation of the illness that caused "non-compliance". Rather the women were primarily concerned with controlling their symptoms and they felt constrained by the demands of the life situations in which they employed treatments. "For these women the issue is not compliance, but controlling symptoms with treatments they can live with" (Hunt, Jordan, Irwin & Browner, 1989, p. 330). This raises the issue of the goal of treatment, whether it should be compliance with medical regimes or optimal adjustment to living with chronic illness. Roberson (1992) suggests nurses need more understanding of client perspectives in order to assist clients to adjust to illness.

Illness - stress, adaptation and coping

A major perspective in nursing (Roy, 1984) and in psychology is to consider illness as a stressor to which the individual must adapt by using coping processes. Illness is one of many stressors a person may cope with in life. "Psychological stress is a particular relationship between the person and environment that is appraised by the person as taxing or exceeding his or her resources and endangering his or her well-being" (Lazarus & Folkman, 1984, p. 19). Psychologists have explored the intrapsychic processes a person uses when coping with stress but there is now an increasing emphasis on considering the environment that people live in. The intrapsychic processes of appraisal and coping have been defined by Lazarus and Folkman (1984). When experiencing stress the person undertakes a "cognitive appraisal", evaluating the stressful situation with respect to its significance for well-being, and what, if anything, can be done about it. Cognitive appraisal is individual and determined by a person's commitments (what is important to a person, what has meaning to him or her) and beliefs, particularly about personal control. Situational factors, including uncertainty and previous experience, also influence this appraisal. Lazarus and Folkman (1984) distinguish between *"coping and automized adaptive behaviours* by limiting coping to demands that are taxing or exceeding a person's resources" (p. 141-142). Therefore a person is coping only when they are experiencing stress. Coping can be emotion focused where the person regulates their own response to the stressor and includes denial, intellectualisation, avoidance, detachment and minimisation. Coping can also be problem focused. There is a seeking to alter the stressor and this may include information seeking, direct action, inhibition of action and using social support. Both types of coping are equally important and are used by most people in stressful situations (Folkman & Lazarus, 1980).

Benner and Wrubel (1989) have built on this view of illness as a stessor and have taken the further step of stating that "as long as one has no symptoms or other disruption of usual functioning, there is no experience of illness

even though disease may be present." (p. 59). We must be conscious of physical, personal or social disruption to experience being ill.

Roy's (1984) adaptation model incorporates intrapsychic adaptive processes but gives equal emphasis to role function, social support and physiological functions as adaptive responses. The stressor (illness) is the focal stimulus but is accompanied by contextual stimuli that alter a person's perception of the impact of illness. These include age, sex, education and health related activities that accompany the illness experience. Residual stimuli are unknown factors that may also influence the situation.

There are no published nursing studies of adults with CF which have used the stress adaptation and coping theoretical framework. Pollock (1993) studied people with other chronic diseases and identified variables that related positively to both physiological and psychosocial adaptation for the total sample: having the "hardiness characteristic", having an ability to tolerate stress, and health promotion activities. There was also evidence that the person's perception of the disability caused by the illness is an important variable in the process of adapting to chronic illness.

Pollock (1989), based on work performed by Kobasa (1979), defines hardiness as comprising three factors: the ability to exercise control, a commitment to be actively involved in life, and an acceptance of challenge which includes a belief that change is beneficial. Pollock (1993, p. 86) states that "adaption to chronic illness is a complex process involving numerous internal and external factors that influence response and subsequent level of adaption".

Schussler (1992) proposes that moderate denial is necessary to adapt to illness, specifically the denial of threat potential or uncertainty. Schussler tested the illness concepts developed by and based on Lipowski's (1970) clinical experience. These are illness as challenge, illness as enemy, illness

as punishment, illness as weakness, illness as relief, illness as strategy, illness as irreparable loss or damage, and illness as value. Lipowski hypothesised that illness concepts are linked with mental well-being. Schussler concludes from his study that these concepts can be linked to the self concept. Illness as challenge

> ...shows the integration of illness in the self concept, whereas illness as damage shows the overwhelming threat of illness in the face of low self esteem. Illness as enemy and punishment are not integrated in a concept of self; the integration between self and illness is based on immature defences (Schussler, 1992, p. 430-1).

Schussler goes on to state that

> ...emotion-related coping modi frequently occurred in persons who did not accept their illness or do not consider it controllable whereas problem-related coping strategies tend to be found in patients who accept their illness or believe they may be able to influence it (Schussler, 1992, p. 430).

He also states that 45% of the patients with rheumatoid arthritis who took part in the study saw their disease as a threat and an enemy and this attitude was related to the severity and the prognostic uncertainty of their disease. Apart from this comment there was little attempt to link the illness concept to disease severity or any other external factors that may influence the person's self concept such as physical functioning, social role or having a restricted life.

Charmaz (1983), when studying suffering in the chronically ill, found that loss of self was caused by leading restricted lives, experiencing social isolation, being discredited and burdening others. She comments on the

nature of suffering, suggesting that when one is suffering there is a language of loss and self-concern; it is only those who have improved and no longer suffer who can see it as a path to knowledge and self discovery.

The impact of illness on the person

There is increasing value placed on exploring the experiences of those who have suffered from chronic illness, particularly if they can put it in context for others as well (Conrad, 1990). Stories of illness reveal the impact that the illness has on the person. The main theme of these narratives is the effect that the changes associated with the disease have on the body and on the self.

> Nothing so concentrates experience and clarifies the central conditions of living as serious illness. The study of the process by which meaning is created in illness brings us into the everyday reality of individuals like ourselves, who must deal with the exigent life circumstances created by suffering, disability, difficult loss, and the threat of death...Illness narratives edify us about how life problems are created controlled and made meaningful (Kleinman, 1988, p. xiii).

Sacks (1984) has written of his experience of breaking his leg and how the subsequent loss of function in this limb affected his whole self.

> I could no longer remember how I had ever walked and climbed. I felt inconceivably cut off from the person who had walked and run, and climbed just five days before. There was only a 'formal' continuity between us. There was a gap - an absolute gap - between then and now; and in that gap, into the void, the former 'I' had vanished (p. 58).

Sacks' story illustrates the concept of humans as embodied beings. When healthy we take our bodies for granted; in illness the body is experienced as other and in opposition to self (Toombs, 1987). Illness is not only experienced as a threat to the body but as a threat to the self. Loss of certainty, loss of control, and change in perception of the future accompany illness. In illness one's body becomes prominent, and both the continuity of daily life and its concerns are disrupted. Illness has its own demands and is often experienced as being beyond the person's control, requiring the person to surrender autonomy and integrity (Zaner, 1985).

Register (1987) writes that decisions on how much to reveal about her own illness are based on not always wanting to be conscious of her illness or give it too much attention. She also has the desire not to be viewed as ill, which diminishes her as a person. Thus, feeling well in chronic illness is the absence of symptoms; not being aware of one's body.

> We learn to accept, or acknowledge, the seriousness and permanence of our illnesses, and then practice a selective denial. We can live as well as possible only if we ease up on the pervasive consciousness of illness to make room for all the other thoughts and feelings that a full life requires (Register, 1987, p. 197).

Not all sickness threatens personhood, but when it does it is experienced as a loss of self or as suffering (Charmaz, 1983). This is not well understood by health professionals who may focus only on curing or alleviating disease rather than alleviating suffering. Cassell (1982 as quoted in Brody, 1991), states

> ...relief of suffering comes most often by changing the meaning of the experience for the sufferer and restoring the disrupted connectedness of the sufferer with herself and those around

> her. Indeed, modern medical practice, by focusing exclusively on bodily pain and ignoring the multiple aspects of personhood and personal meaning, may inadvertently increase suffering while seeking to relieve it (p. 30).

Lieb (1976) discusses concepts of health and concludes that health itself is of no value; it is the fullness of life with which we have to be most concerned. Health allows us to live our lives and attend to the concerns which define us. In illness these concerns remain the same. He discusses how, when we are ill, we are to act as we act when we are well, though perhaps less strenuously. When we are dying we have the same concerns as before, we are our concerns.

Brody (1991) used literary works with stories of sickness to identify the effects on the person. He explores the dual nature of sickness, "...the way it can make us different persons while we still remain the same person" (p. x). To be sick is to have something wrong with oneself, to experience disruption of the body and self and a threat to one's integrated personhood. Illness is accompanied by alteration in social roles, and disruption in biological, social and cultural systems.

Brody suggests that patients have to work out meanings and life plans for themselves, and that physicians can assist this process both by understanding patients' stories of the illness and by being aware of the power of physicians' stories. The physician can relieve a patient's suffering if he "...explicitly recognises the relationship between sickness and self respect, and carefully looks for clues as to the link between various aspects of the illness, the reaction of the social peer group to the illness, and the patient's previous and future life plans" (p. 191).

There are no "patient perspectives" of adults with CF in the literature. However, there has been some exploration of the psychosocial response of

adults with the disease and these will be discussed in the following section. These studies have not attempted to uncover the lived experience of adults with CF.

CYSTIC FIBROSIS (CF)

CF is like other chronic illnesses in many respects, but there are variations in how different illnesses are experienced because of their symptoms and the social meaning of the disease. For example, Williams (1993) found that people with Chronic Obstructive Airways Disease (COAD) experience the distress associated with dyspnoea, and this appears to be affected by anxiety. This was not revealed in this study of people with CF. People with COAD are also of a generally older age group so they experience different biographical and social concerns. People with AIDS who experience respiratory symptoms are generally of a similar age group to adults with CF, but the social stigma and guilt related to their illness are much more pronounced (Weitz, 1989) than in people with CF who have "nothing to hide and nothing to advertise" (Admi, 1995).

Incidence

The incidence of CF in Caucasian populations is estimated at 1:2500 live births (Tsang, Hodson & Yacoub, 1992). It does occur in other population groups but to a much lesser extent. Until the 1950s, most children with CF died in the first year of life. Improvements in the understanding and treatment of the disease have gradually led to an increased life expectancy and thus, more adults with CF. In 1990, 36% of people with CF in England and Wales were over 16 years of age (Smith & Stableforth, 1992). Information collected on the New Zealand National Cystic Fibrosis (CF) database reveals that 94 people out of a total of 302 are older than 15 years (Wesley, Dawson, Hewitt & Kerr, 1993). In the United Kingdom, the number of affected adults is expected to increase steadily in the foreseeable future because of improvements in medical care. Over 50% of people with

CF are expected to live to the age of 30 years by the year 2000 (Tsang et al., 1992). Improvement in life expectancy can also be expected in New Zealand, although projected survival rates are less than some overseas centres (see Introduction). Wesley and Stewart (1985) published a report of the incidence and mortality of the disease in New Zealand. They found that New Zealand has a lower incidence than the United Kingdom perhaps because of delayed diagnosis and possibly because of racial mixing in the population.

Aetiology

Super (1992) discusses the history of the knowledge about CF. According to European folklore a child with a sweaty forehead would die young. The first clear clinical descriptions of the disease occurred in the 1930s. Knowledge about CF has been expanding over the last sixty years.

CF is now known to be inherited through an autosomal recessive gene. Both parents must be carriers of the gene defect for a child to have the disease. In 1989 the CF gene was localised. Multiple mutations of the gene have been identified. This may explain the variety in the type and severity of symptoms experienced by different individuals, but a common gene defect accounts for 70% of cases (Cleghorn & Isles, 1992).

Pathophysiology

For many years the pathophysiology of the disease could not be completely explained. It is now thought that the gene defect effects the transport of chloride ions by the epithelial cells of exocrine glands causing their secretions to be thicker and "stickier" than normal. This interferes with normal lung function and makes the person more vulnerable to infection. The upper and lower airways, the pancreas, gastrointestinal and reproductive systems are affected. People with the disease usually develop progressive respiratory disease following repeated chest infections. Most have abnormal pancreatic function because the pancreatic ducts are blocked

preventing secretion of digestive enzymes and leading to fat and protein malabsorption. Diabetes mellitus can also result from pancreatic obstruction. Nearly all males with the disease are infertile due to obstruction of the vas deferens, the seminal vesicles and the epididymis. Females have reduced fertility because of thickened secretions. Respiratory infection or deterioration in lung function is the main reason for hospitalisation, and the leading cause of mortality. There is enormous variation in the pattern and severity of symptoms.

Diagnosis

Before the introduction of neonatal screening infants with CF presented with a variety of symptoms: meconium ileus, recurrent chest infections, malabsorption, failure to thrive, offensive bulky stools, rectal prolapse or obstructive jaundice (Russell, 1978). In the past, mild symptoms meant some patients were not diagnosed until over 15 years of age (di Sant'Agnese & Davis, 1979). Since the introduction of neonatal screening the average age of diagnosis is now six weeks. Increased immunoreactive trypsin is detected as part of a routine neonatal blood spot test (Ryley, Goodchild & Dodge, 1992). Diagnosis is still confirmed using the sweat test which identifies elevated chlorine. Earlier diagnosis is expected to lead to better health and prolonged life expectancy. Prenatal diagnosis and identification of carriers is now possible but is confined to families with a known history of the disease. There are ethical, financial and resources issues surrounding population screening that are presently being debated (Friend, 1990; Fernbach & Thomson, 1992).

Respiratory symptoms and management

Respiratory disease is the major cause of morbidity and eventual mortality in CF. A survey of adolescents and adults found that respiratory infection was responsible for 90% of morbidity and mortality (Penketh, Wise, Mears, Hodson & Batten, 1987). Respiratory disease accounted for 97% of deaths and 75% of admissions. Neonates are thought to be born with normal lungs.

An initial infection is followed by hypersecretion and airway obstruction with thick mucus that causes susceptibility to further infection. Chronic colonisation by bacterial pathogens leads to acute exacerbations of progressive lung disease. This colonisation follows a sequence over the person's life: Staphyloccocus *aureus* in infancy, Haemophillus *influenzae* during the early years, Pseudomonas *aeruginosa* in adolescence and Pseudomonas *cepacia* and mycobacteria during the late teens (Hodson & Warner, 1992).

Daily physiotherapy and regular physical exercise are recommended for all people with CF in order to clear secretions. Prophylactic antibiotics are used and lung infections are treated aggressively. Pseudomonas *aeruginosa* can be treated with oral ciprofloxacin but, in New Zealand, if there is no improvement, the person will be hospitalised for intravenous antibiotics. Intravenous antibiotic treatment at home has been developed overseas for both children and adults with CF but it is not the norm in New Zealand (Kendrick, 1993). Home treatment is reported to have many benefits: reduced hospital admissions, less disruption to schooling and employment, decreased cross infection, earlier treatment and cost effectiveness (Catchpole, 1989). Decreased length of stay in hospital, improved lung function and quality of life have also been found as a result of using a "Intermate" system of home-care intravenous antibiotics (Bramwell, Halpin, Duncan-Skingle, Hodson & Geddes, 1995). Sinusitis is almost always present in adults with CF (Hodson & Warner, 1992).

Repeated lung infections lead to scarring of the airway, reduced lung function and eventually hypoxia, pulmonary hypertension with cor pulmonale in the end stage. Pneumothorax and pulmonary haemorrhage are possible developments with progressive lung disease. Heart-lung and lung transplants have been successfully performed in the United Kingdom since 1985. The scarcity of donors and the criteria for selection mean that it

is an option for only a minority of adolescents and adults (Tsang et al., 1992).

Gastrointestinal symptoms and management

Malabsorption occurs as a result of the poor digestion of food secondary to pancreatic insufficiency. Enzyme therapy is essential. Other potential complications are cirrhosis of the liver, diabetes mellitus, and distal intestinal obstruction syndrome. As recently as the late 1970s, a low fat diet with artificial preparations was advocated by some clinicians (Russell, 1978). People with CF used to have chronic undernutrition initially due to malabsorption, and then later due to their energy needs not being met on a low fat or a normal diet. They are now encouraged to increase their dietary fat and have high calorie snacks between meals - 120-150% of the normal calorie intake is recommended. Short term gastrostomy supplemental feeding is advocated for those with advanced lung disease who are unable to maintain a positive energy intake because malnutrition may contribute to the progressively deterioration of lung function (Cleghorn & Isles, 1992).

Improving life quality and expectancy

Improvements in management and life expectancy resulted from the discovery of B-lactamase resistant penicillins and pancreatic extracts (Super, 1992). The advent of specific CF organisations has led to more research and better care. "The cornerstones of CF medical treatment remain optimised nutrition, antibiotics and chest physiotherapy" (Alton, Caplen, Geddes, & Williamson, 1992, p. 785). New treatments such as nebulised amiloride and DNase are being trialed. Several lung transplants have been carried out in the United Kingdom with success, and this could possibly happen in New Zealand. As the gene defect is now known, gene therapy appears likely in the future. Spraying the lungs with the healthy counterpart of the CF gene may correct the defect, but this treatment is still several years away (Smith & Stableforth, 1992).

Recent nursing studies have examined the nurse's role in the care of people with CF and their families. Whyte (1992) has studied the contribution of nursing to the support of the family. She examines the difficulties that parents face, and discusses nursing involvement within a family nursing framework. Two articles have looked at the issues surrounding the administration of intravenous treatment at home and the benefits it can bring to people and families as their lives are less disrupted by this than by hospitalisation (Catchpole, 1985; Gill, 1993). Two articles discuss the role and contribution to care of the CF specialist nurse within a paediatric context in the United Kingdom (Glew, 1993; Moore, 1988).

Psychosocial impact

The psychosocial impact of CF has been examined by nursing and medical researchers. Most nursing studies of CF have focused on parents who are coping with a child with CF, and the nurse's role in educating and supporting families. The stress that the illness and the treatment regime places on the family and parental coping strategies have been examined (McCubbin, 1984; Gibson, 1986). Canam (1986) has examined the information that parents need to facilitate communication about the illness within the family. Van Os, Clark, Turner and Herbst (1985) sought to examine the relationship between life stress in families and illness exacerbation. Patterson, McCubbin and Warwick (1990) examined the effects that family function has on indices of health for the child with CF. These studies all emphasise that the ill child cannot be treated in isolation from their psychosocial environment, and family functioning must also be considered.

There have been few nursing studies that have focused on adults with CF and how they live with the disease. Dibble and Savedra (1988) used a case study to illustrate the management of CF in adolescence. Baker and Coe (1993) used their own practical experience and Havighurst's (1969) Theory of Developmental Tasks to write about the nurse's role in assisting the

transition to young adulthood. A study by Brissette, Zinman and Reidy (1988) reveals that a group of young adults disclosed their concerns in a pattern that reflected their increasing trust with the nurse involved in their care. Initially they would talk about CF management, then growth and development issues. After a year they had built up enough trust so that they could talk about family relations. Brisette, Zinman, Fielding and Reidy (1987) developed a care plan based on these findings. A recent study by Admi (1995;1996), using a grounded theory approach, found that young people with CF are very similar to their peers in the way they lead their lives. Having a chronic illness was not central to their view of themselves nor did it determine how they lived their lives.

By the late 1970s there were sufficient numbers of adolescents and young adults surviving with CF for medical researchers to begin to study the social functioning and psychosocial adaptation of these age groups. di Sant'Agnese and Davis (1979) reviewed 75 cases and another 232 cases in the literature, and concluded that the majority of adults with CF achieve academic success, are involved in employment and form partnerships. This study alluded to difficulties such people face related to their illness but did not explore them in any depth.

Most other studies have attempted to measure social functioning and psychological effects, mainly through the use of questionnaires and psychological testing. Boyle, di Sant'Agnese, Sack, Millican, & Kulczycki (1976), through psychiatric interviews and psychological testing, concluded that the 27 adolescents and young adults who took part in their study appeared to function on a day-by-day basis. They found various levels of psychological distress resulting from the condition, with the prime causes being altered physical appearance, strained interpersonal relationships, conflicts in upbringing and increased awareness of the future and of death. A psychosocial function questionnaire administered to twenty one patients by Strauss and Wellich (1981) confirmed these findings: participants were

found to experience psychological distress about appearance and social isolation, and used denial and minimisation as coping mechanisms. The study also found adequate social and occupational success in the majority of participants in the study.

Simmons et al. (1985) studied a group of 12-15 year olds and found them to be functioning well with the majority having a good self concept. Pinkerton, Trauer, Duncan, Hodson and Batten (1985) found considerable variation in the individual adult's ability to cope with the disease regardless of the severity. The group they identified as non-copers had a lack of health care knowledge leading to non-compliance, and their families had a lower level of effective functioning than the others. Shepherd et al (1990) undertook a comparative questionnaire of 37 adults with 46 healthy peers and concluded that there was little difference between the two groups for measures of emotional support, social network density, self esteem or current life satisfaction. Walters, Britton and Hodson (1993) found a high proportion of CF adults are living full and productive lives.

Aspin (1991) reviewed the findings of psychological research about adults with CF since 1979. He concluded that there is evidence of psychological distress among adults but none of these studies (Boyle et al., 1976; Bywater, 1981; Cowen et al., 1984; Moise, Drotar, Doershuk & Stern, 1987) had identified or classified it. Aspin also discussed studies (Moise et al., 1987; Shepherd et al., 1990; Strauss & Wellich, 1981) in relation to attention and avoidance strategies and concluded that they shows that people with CF fare better when using avoidance.

Conclusion

The literature on chronic illness reveals that illness not only affects the person physically but also psychosocially. At times these concerns may outweigh physical ones. Living with a chronic illness requires adjustment by the individual and family, and a chronic illness can be described as

having a trajectory with different phases. There is work required to look after the illness and its biographical impact. A person with a chronic illness is not usually a passive recipient of health care as is more likely to be the case in an acute facility. Instead they are likely to be actively involved in the management of their illness. Illness can be seen as a stressor with which the person must learn to adapt, using coping strategies which may be intrapsychic or external. Illness differs from the disease process and is dependent on a person's perception of disruption. Illness can also affect personhood. Its effects on one's body can also mean the self is also profoundly affected.

The literature on CF has mainly focused on the management of the disease. Several studies have attempted to measure the effects of CF on adolescents and adults. Some studies have found that the majority of adults with CF are leading full and active lives.

CHAPTER TWO
RESEARCH METHOD

The purpose of this study is to increase understanding of an illness as it is realised in the everyday life of adults with CF. Hermeneutic phenomenology was chosen as the research method because its philosophical basis is consistent with nursing's desire to understand the nature of particular human experiences. This chapter discusses the philosophical assumptions behind hermeneutic phenomenology, describes phenomenological inquiry and the limitations of phenomenological research. The design and method of the study including ethical considerations and information about the study participants are then outlined, and issues relating to the validity of phenomenological research are discussed. An overview of the findings completes the chapter.

ASSUMPTIONS OF PHENOMENOLOGICAL RESEARCH

Phenomenology as philosophy seeks to increase understanding of human phenomena. It differs from the scientific tradition in that it does not seek to develop knowledge based on proven fact. Edmund Husserl (1859-1932) sought to "establish a science of phenomena as the cognition of *essences* rather than of matters of fact" (Annells, 1996, p. 706). He sought to change the philosophical thinking of his time by emphasising subjective rather than objective knowledge. Husserl's student, Martin Heidegger (1962), developed another distinct branch of phenomenology concerned with the nature of being and of understanding what it means to be a person.

Heideggerian phenomenology assumes that things have significance and value to persons; that a person is his or her concerns. One of the things that makes us human is that we care, that things matter to us. "Each one of us is what he pursues and cares for. In everyday terms we understand ourselves and our existence by way of the activities we pursue and the things we take

care of" (Heidegger, 1962, p. 159). Concerns differ between individuals due to differences in culture, beliefs and life situations. "To understand a person's behaviour or expressions, one has to study the person in context, for it is only there that what a person values or finds significant is visible" (Leonard, 1989, p. 46).

Heidegger's exploration of the nature of being led him to a concept of "Dasein" or *being-in-the-world* where each person has a world or *lifeworld.* This is not the person's physical environment but "the meaningful set of relationships, practices, and language we have by virtue of being born into a culture" (Leonard, 1989, p. 43). Meanings and everyday practices of the *lifeworld* shape the person and set up possibilities as to who a person can and cannot become. Persons are situated and cannot be separated from or free of the language, culture, history, purposes, and values that they have been shaped by, and that surround them. Persons are not conscious of the *lifeworld*; it is normally taken for granted. "World is so all pervasive as to be overlooked by persons, and it only appears to us in a conscious way when disruption or breakdown occur" (Leonard, 1989, p. 45).

An assumption of Heideggarian phenomenology is that persons are self interpreting; that they understand themselves and others by interpreting events, emotions and behaviours according to the beliefs and cultural values of their *lifeworld.* Self interpretation occurs largely unselfconsciously; a person is not aware of consciously thinking or theorising about everyday practices and behaviours.

Phenomenology also assumes that it is through our bodies that we access the world. "In the phenomenologic view, rather than having a body, we are embodied. Our common practices are based on shared embodied, perceptual capacities" (Leonard, 1989, p. 48). Normally we take our bodies for granted and are not conscious of them. This breaks down in illness and we become aware of the "taken-for-granted understanding of health: the unity of self

and healthy body" (Leonard, 1989, p. 48). Bodily breakdown disrupts our ability to negotiate the world.

Temporality is also an important concept in phenomenology. Time is not linear but viewed as connectedness (Annells, 1996). The person or *Being-in-time* cannot be separated from the past or from his or her projected future.

PHENOMENOLOGICAL INQUIRY

Phenomenological inquiry seeks to increase understanding of what it means to be human by studying human experiences in their context and complexity. The world of everyday lived experience is captured and described. "Understanding of the person cannot occur in isolation from the person's world" (Walters, 1995, p. 792). Everyday practices and assumptions usually remain in the background and these are what phenomenology seeks to reveal.

> Phenomenology aims at gaining a deeper understanding of the nature or meaning of our everyday experiences. Phenomenology asks, 'What is this or that kind of experience like?' It differs from almost every other science in that it attempts to gain insightful descriptions of the way we experience the world pre-reflectively, without taxonomising, classifying or abstracting it. So phenomenology does not offer us the possibility of effective theory with which we can now explain and/or control the world, but rather it offers us the possibility of plausible insights that bring us into more direct contact with the world (van Manen, 1990, p.9).

The method of phenomenology is to collect descriptions of a human phenomenon from those who are living the experience and know it best. These descriptions are then interpreted by the researcher. "Understanding

human action always involves an interpretation, by the researcher, of the interpretations being made by those persons being studied. This interpretive approach is called hermeneutics" (Leonard, 1989, p. 50). The interpretation seeks to uncover themes or concepts that capture the essence of the phenomena being studied. Heiddegger (1962) argues that it is only possible to interpret something according to one's own lived experience. There are no interpretation free facts for phenomenologists to describe. The researcher therefore participates in a hermeneutic circle (Reinharz, 1983).

> In interpreting text one must move back and forth between an overall interpretation and the details that a given reading lets stand out as significant. Since the details can modify the overall interpretation, which can in turn reveal new details as significant, the circle is supposed to lead to a richer and richer understanding of the text" (Dreyfus, 1991, p. 36).

There must be constant reference between the part and the whole of the text to maintain integrity. In this way the researcher is challenged to stay true to the text and honour the lived experience of the participants.

Phenomenological inquiry is dependent upon the relationship between researcher and participant, and the researcher's ability to engage with their reality; the researcher needs empathy, intuition and attention, and also some understanding of the participant's experience. The researcher's beliefs and experience contribute to the interpretation process. In Heideggerian phenomenology there is no attempt to bracket or exclude the experiences, bias or prejudices of the researcher as with Husserlian phenomenology. Rather they are considered to be an important part of the research process (Walters, 1995).

Hermeneutic phenomenology is being increasingly used in nursing research as it is suited to the study of many nursing phenomena (Annells, 1996).

Nursing is concerned with "the phenomenon of health and the human predicament of coping with illness" (Watson, 1979). Descriptions of these gained from a phenomenological perspective will lead to greater understanding of the illness experience and increase nurses' ability to assist people as they live their lives.

LIMITATIONS OF PHENOMENOLOGICAL RESEARCH

The purpose of phenomenological research is to explore and suggest emergent themes rather than to make definitive statements or establish a generalisable theory. The finding of a phenomenological study can only be used to increase understanding but not be held up as being absolute fact.

Phenomenology is only useful for studying subjective phenomenon. It relies on the participants to reveal an experience and then the researcher to interpret those experiences. It is not suitable for situations where the participant is unable to describe the experience or when the researcher cannot truly understand it. For example, cultural difference between participants and researcher may preclude understanding of aspects of a phenomenon.

DESCRIPTION OF THE RESEARCH PROCESS

The experience of adults living with CF interested me because of the gap I perceived between the nursing services being provided to this group and what I thought might be more relevant to their health needs. There was the potential to enhance the relevance of nursing if we had a greater understanding of that experience. To investigate the phenomenon of living with CF, four adults with the disease were invited to describe their experiences. Initially, I thought that I may want to interview partners or family members, but it quickly became obvious that this would be beyond the scope of this study. The number of participants is small and my

intention was not to represent the variety and variability of this experience but to provide a depth of understanding. The interviews were unstructured and ranged in length from half an hour to two and a half hours. Two of the participants I had previously interviewed as part of a previous fieldwork so there was an established rapport. I interviewed them both two times and had several telephone conversations with one. Two other participants I interviewed only once. For Lincoln and Guba (1985), the degree of structure in the interview is dependent on who most appropriately holds the questions:

> ...the structured interview is the mode of choice when the interviewer *knows what he or she does not know* and can therefore frame appropriate questions to find it out, while the unstructured interview is the mode of choice when the interviewer *does not know what he or she does not know* and must therefore rely on the respondent to tell him or her (Lincoln & Guba, 1995, p. 269).

Early interviews began with broad, open questions. For example: "Tell me what it is like for you living with CF". As data collection and analysis of interviews was concurrent, some participant interviews had more focused questions.

Van Manen (1990) identifies six research activities inherent in the phenomenological method. These are:

1. Turning to a phenomenon which seriously interests us and commits us to the world;
2. Investigating experience as we live it rather than as we conceptualise it;
3. Reflecting on the essential themes which characterise the phenomenon;

4. Describing the phenomenon through the art of writing and rewriting;
5. Maintaining a strong and oriented [pedagogical] relation to the phenomenon;
6. Balancing the research context by considering parts and whole

(van Manen, 1990, p. 30-31).

These guiding steps have been followed in this study. The nature of living with an illness is important to me and relevant to my work as a nurse. Interviewing participants who were living with the phenomenon is the closest way of investigating subjective experience. I transcribed the interviews and then read and reread them, reflecting on parts and then the whole. I looked for similarities, and complementary and conflicting points of view, among the participant interviews. I identified themes that I believe reflected the participants' experiences. Themes are described by van Manen (1990) as "the form of capturing the phenomenon one tries to understand" and "the means of getting at the notion"(p. 87-88). Relevant literature was explored for evidence of similar themes and experiences to those that were emerging in this study.

Themes were developed from phrases of the participants that captured the essence of their experience. Sometimes a word or phrases (or a variation) was repeated by one or more participants. Dictionaries were consulted to clarify the meaning and use of various words. Some themes or similar themes were found in the literature. The word "effort" was used rather than "work", which is a concept that has already been developed in the chronic illness literature (Strauss & Glaser, 1975; Corbin & Strauss, 1988). "Work" was never used by the participants whereas effort was, and it implied a more conscious response to a demand of the illness on their part.

Van Manen (1990) emphasises the importance of writing as part of the hermeneutic process. It is not the final step before presenting the study, but integral to the research process. In this study the development of emergent themes has been accompanied by memoing, identifying illustrative examples and then rewriting to clarify and make the meaning obvious. Throughout this process parts have been constantly considered in the light of the whole. My conscious goal has been to present the outcome in a way that captures the essence of the participants' experience.

THE PARTICIPANTS

All four participants were over the age of eighteen. There was no attempt to select on the basis of gender, lifestyle or disease severity. Participants were contacted through the Cystic Fibrosis (CF) Association. They were asked by a member if they would be willing to participate in the research before being approached by me. A copy of the information sheet and consent form are included in the Appendices.

The commitment to protect participants' anonymity means that the four participants will be described as a group rather than individually. They are all within the age group 20 - 40 years. Both sexes are represented. There is variety in both the symptoms and severity of the disease. Two of the participants feel relatively well while the other two are experiencing some instability in their condition. Two of the participants were diagnosed in early childhood, the other two were older. One of the participants is employed. Two are in stable partnerships, and one has a child.

ETHICAL CONSIDERATIONS

Permission for the study was obtained from the Massey University Human Ethics Committee and the Central Regional Health Authority Wellington Ethics Committee. Ethical considerations influenced the design of this

study. To prevent any discomfort or potential risk, participants were only interviewed when well. This meant that people in a very progressive phase of the illness were excluded from the study. The small number of adults with CF in New Zealand means there are difficulties maintaining the confidentiality and anonymity of participants. This was explained before they gave consent to participate. I undertook to take every step possible to maintain their anonymity. This factor also determined that interviews would take place in their own home rather than in a health care facility and excluded participant observation.

VALIDITY IN PHENOMENOLOGICAL RESEARCH

Validity in phenomenological research is not about trying to give data the status of fact or objective truth independent of interpretation. "Heideggerian hermeneutic phenomenology is oriented towards interpretation and understanding, and recognises that there is no such thing as an uninterpreted fact" (Walters, 1995, p. 795). The validity of the study and the researcher's interpretation must be judged by the readers using information about the research process and excerpts from the participants' narratives. Leonard (1994) and Benner (1994) both support and expand on this contention.

> A study can be judged on how carefully is the question framed and initial interpretive stance laid out, how carefully the data collection is accomplished and documented, and how rigorous the interpretive effort goes beyond publicly available understandings of a problem to reveal new and deeper possibilities for understanding (Leonard, 1994, p. 61).
>
> . . . . . .
>
> The interpretation must be auditable and plausible, must offer increased understanding and must articulate the practices, meanings, concerns and practical knowledge of the world it

> interprets. Good interpretation is guided by an ethic of understanding and responsiveness. One must not read into the text what is not there. Self knowledge is required to limit the interpreter's projection of his or her world into the text (Benner, 1994, p. xvii).

The interpretation remains tentative rather than absolute or true. By providing excerpts or illustrations, the validity of interpretation can be judged by readers (Walters, 1995).

OVERVIEW OF THE FINDINGS

This chapter has briefly outlined the assumptions behind phenomenology and described the research method. The next four chapters will present the findings of this study and describe the experience of adults living with CF. The findings will be presented using themes which capture the meaning of this experience and increase our understanding of it.

In Chapter Four *realising CF* by the participants will be described. Although the participants have lived with CF from birth they do not always experience themselves as having an illness. Some experienced *realising the meaning of CF* as teenagers. Realising involves gaining knowledge about the disease but also involves experiencing a deterioration in their physical condition; **their** CF was "conceived as real" and the meaning of **their** disease was realised. Changes in their physical conditions confirmed that the general progress of the disease they had learnt about was now true for themselves. Realising is not described as a sudden event but a gradual awareness of the implications of having the disease. *Realising CF in everyday life* describes how the extent of the participants' awareness is determined by the phase of their illness. During exacerbations they are much more aware of CF; when experiencing periods of wellness, although signs and symptoms of CF are experienced, they are not so aware of them.

In Chapter Five *learning to live with CF* includes *learning limits* and *making a conscious effort to look after themselves. Learning to manage* includes *learning to manage illness*, *learning to manage hospitalisation* and *learning to manage relationships with health professionals.*

In Chapter Six the importance the participants placed on *getting on with life* rather than focusing on their CF is described. The desire to get on with life means people with CF are *balancing treatment regimes, minimising illness to self and to others*, and *rejecting the patient role.*

In Chapter Seven *fighting to live with CF* is described. The strategies the participants use when they experience their health deteriorating and they have to work harder at maintaining their well-being, are described. *Redefining what is important, envisaging a future* and *fighting to maintain a positive attitude.* Those who experience their health deteriorating have to work harder to get on with life, and to stop the disease impinging on their other concerns. They experience *fighting against CF.*

These themes are summarised on Table 1.

CHAPTER FOUR

REALISING CF

Realising the meaning of CF

Realising CF in everyday life

CHAPTER FIVE

LEARNING TO LIVE WITH CF

Learning to live

Learning limits

Making a conscious effort to look after self

Learning to manage

Learning to manage illness

Learning to manage hospitalisation

Learning to manage relationships with health professionals

CHAPTER SIX

GETTING ON WITH LIFE

Balancing treatment regimes

Minimising illness to self and to others

Rejecting the patient role

CHAPTER SEVEN

FIGHTING TO GET ON WITH LIFE

Fighting against CF

Redefining what is important

Envisaging a future

Fighting to maintain a positive attitude

Table 1. Summary of Themes

CHAPTER THREE
REALISING CF

I suppose just realising that...that's when I saw that I wasn't going to be as big as everybody else. Realising, it was sort of a realisation of cystic fibrosis (CF). It's hard to explain.

This chapter will discuss how people with CF come to a realisation of CF. Although the disease has been present from birth it is not until the disease is understood, and not until they feel significantly affected by CF, that they realise the personal meaning of the disease. They realise, or are aware, of CF in everyday life but the extent of this realisation is dependent on how much the disease is affecting their health. There are two themes associated with realising CF: *realising the meaning of CF* and *realising CF in everyday life.*

REALISING THE MEANING OF CF

Dimond and Jones (1983) make a useful distinction between disease and illness. **Disease** is "a state in which the body is suffering from a malfunction in one or more parts" (p. 3). It is the course of the *biological* process including the signs (observable indicators such as rash or fever) and symptoms (unobservable indicators such as pain or nausea). **Illness** is the "altered perception of the ill person" and includes the symptom experience. "Disease can exist in the absence of illness" (p. 5); a person may have a disease without being aware of it, without experiencing any signs or symptoms of the biological process.

CF is a hereditary disease. People may know that they have it all their lives. They may be actively involved in treatment and preventive measures from an early age including taking enzyme tablets with meals and having frequent chest physiotherapy. But knowing they have the disease and

participating in treatments does not necessarily mean the person experiences himself as being ill or as having an illness. One of the participants describes his growing realisation of his illness:

> *When I was younger, I didn't really have much concept of what it was. I knew I had CF. By the time I went to school I was taking pills, swallowing capsules. But I never felt that physically affected by it until I was about 17. Then I started, like my health went down a little, and it's just becoming progressively worse. So now it's a really big thing in my mind. Sort of think about it when you wake up every morning. So I feel much more affected now than I did years ago...I suppose just realising that sort of...that's when I saw that I wasn't going to be as big as everybody else. Realising , it was sort of a realisation of cystic fibrosis (CF). It's hard to explain.*

This participant, who was diagnosed in infancy, did not feel significantly affected by CF until he perceived that his "health" had deteriorated, and that he was smaller than others in his age group. Changes within his body made him aware of CF. Until this time he had experienced his "disease" not as physical symptoms but as the outward signs, namely, the tablets and the physiotherapy. It was not until the disease affected him in a way that disrupted his "taken-for-granted understanding of health: the unity of self and healthy body" (Leonard, 1989, p. 48) that he become conscious of having an illness.

In the phenomenological view of what it means to be a person, we are embodied; our selves (our minds) are not separate from our bodies. Usually we are not conscious of our bodies, we take them for granted and only become aware of them in breakdown or illness. In illness

> ...one's body is no longer the silent means for acting, walking, talking, reaching, seeing, hearing, etc., but becomes prominent. When my eyes hurt, I become aware of my embodied seeing; when my breathing becomes laboured, I am forced to an awareness of it quite unlike my ordinary experience (Zaner, 1985, p. 80-81).

It is then that we become conscious of our bodies and are forced to pay them attention. Health, according to this perspective, is that time when we are unaware of our bodies and hence of our illness. Baron (1985, p.609) describes health "as a state of unselfconscious being that illness shatters".

The same participant also describes starting to realise the **meaning** of his disease. He started to understanding its significance for himself and his life.

> *I suppose being a child and not having a more scientific knowledge of the disease and the way it works...I don't really think [my parents] had any more concept of it than I did. I think I discovered it more for myself than I did from my parents. When I was 17, it was about then that I started reading the literature, books about it and how it works and the genetic side of things. So I could see it from a doctor's point of view as well as a patient's...sort of accepting it and realising that it was actually quite serious and it's not going to go away and it's progressive...It's just going to get worse as the years go by.*

He began to realise that CF was starting to affect his health, that it is a progressive disease, and that he does have a shortened life expectancy. To realise CF in this instance is "to conceive as real; apprehend with the clearness or detail of reality" (The New Shorter Oxford Dictionary, 1993).

Realisation did not happen all at once but rather as he gathered more information and began to understand the physical course of his personal disease.

The meaning of having CF is not understood by children. Admi (1996), when exploring how young people experience growing up with CF, found that

> ...in recalling their experiences as young children, the informants stressed that they did not have an accurate understanding of their disease. They complied with their treatments because they were told to by physicians and parents...around early adolescence (approximately ages 10 and 13) the informants reported more understanding and interest in their disease" (p. 177).

It is not until they gain an understanding about the scientific or medical nature of the disease they can now project how the disease will probably affect them in the future. However, they may not accept what this really **means** or conceive it as real until they begin to personally experience sufficient physical changes that signal that **their** disease is progressive.

Healthy young adults with CF who do not appear to accept their projected lifespan limitations could be interpreted as being in denial. A phenomenological interpretation would argue that healthy adults cannot conceive this as real when they have not yet experienced any change in their embodied self; they have not experienced the loss of their taken-for-granted bodies.

If the disease is diagnosed when people are teenagers or adults and their "health" is already affected, they may comprehend the scientific knowledge immediately and the true meaning of the diagnosis is quickly evident. In

this case, the reaction to diagnosis would be expected to range "from shock and disbelief - 'not my body,' 'not me' (Rosenberg, 1980, p. 49) - to relief that a diagnosis has finally been arrived at" (Corbin & Strauss, 1988, p. 29). Diagnosis was described as unexpected by one participant.

> *I don't feel as though my introduction to CF has been typical at all. It was something that hit me in the back of the head really quite late in life. I almost thought that there are things that can happen to you later on. You can get asthma much later than you ought to. I certainly never felt that I was in line for some kind of hereditary disease. If I thought anything, like that's well behind me, plenty of other things could happen. It was quite unexpected really.*

Another participant describes being diagnosed as a teenager.

> *It was a very strange time especially finding out so late. It filled in a few gaps and it made a whole lot more questions come out of it. To Mum and Dad it was just traumatic, the worst thing that could ever happen.*

This participant reveals the shock experienced by his parents and the need to fill in information gaps on his part. "'Filling in' attempts to answer the question how much, how far and what does 'it' mean?" (Corbin & Strauss, 1988, p. 30). The *personal meaning of CF* is realised as information is gained and understood. The loss of the taken-for-granted body that causes the person to realise the personal meaning of CF also makes the person conscious of CF in their everyday life.

REALISING CF IN EVERYDAY LIFE

To realise also means "to become conscious or aware of (something)" (Collins English Dictionary, 1991). All the participants described becoming and then being aware of CF in their everyday lives. The extent of their awareness was determined by the phase of their illness, whether they were experiencing a period of instability or relative wellness. During periods of wellness, although they had symptoms of CF they did not give much attention to these.

> *I'm always chesty in the mornings. So I just wake up, clear it, and then I'm normally right for the day. But that's something I have to do every day. I'm just so used to it, I don't even really think about it anymore. It's just something I had to do so I did it. If I don't do it though, I know I can feel it for the rest of the... it hurts when I get up. Moving around it clears itself. It's nothing really that I have to think about in the morning, like I have to do it or anything. I'm just used to it.*

For this participant, acute exacerbations of the disease occurred during his childhood. These periods of unwellness may mean that he cannot remember a "taken-for-granted body" because the symptoms of illness have always been part of his experience. The person gets used to experiencing certain symptoms and these become taken for granted.

Another participant who was relatively well at the time describes her awareness of CF in every day life.

> *I don't think there would not be a day go by where I wouldn't think "Oh, there's that little niggly thing there and I know that's cystic fibrosis (CF) there doing that". Everyday there's some little thing. But it might only be just a few moments, or it*

might be a whole day if I've got a bad sinus or whatever, but everyday there's something. Some days it might be as little as taking your enzymes and every time you take an enzyme you think "I can't find those damn enzymes again. Bloody cystic fibrosis (CF)".

There are daily reminders that she has the disease; either minor physical symptoms or treatments, but they are not enough to prevent her from living her life. She has faith and confidence in her body.

I think your confidence in yourself or your confidence in your ability to do things or to be independent... I think your confidence in your independence gets a bit of a shake up every now again. I certainly don't like to be dependent on drugs or too dependent on having people around me who know what to do in a given situation. Your confidence in your ability to carry on doing things or certain things for any amount of time... I think you need a bit of confidence to be able to even go to work. Just to be confident to be able to work full time without physically not being able to handle it.

Life can be interrupted by exacerbations of the illness but, as long as there is a return to their previous state, the participants' confidence in themselves remains.

When I first come out, it sort of takes a bit more. 'Cause when I first come out of hospital I'm lazy 'cause before I went in I was just lazing around. Then, in there you don't do much. Come out, you've got to start exercising again. And it'll probably be another week or some more before you start getting back into your normal routine.
[But you get back to where you were before?]

Yeah I never really go backwards.

This participant has experienced many episodes of hospitalisation but they are not particularly significant to him because he believes that he always returns to his previous physical state. This contrasts with another participant who has experienced a decline in his physical state after his last hospitalisation.

> *This time when I came out it was much more on a level. I didn't feel as weak when I came out but I didn't get back up as high. It seemed much more mundane and in a way much more depressing. I got better but I didn't get significantly better like I did the last time.*

This was an important experience for this man because his health clearly did not return to where it was before the illness episode. During exacerbations of the illness and especially when the disease is being experienced as progressive, the person is much more conscious of his or her state of health and of CF. The person thinks about it more often, and is more aware of the disease affecting self and life, and of the need to give it more attention.

> *Every morning when I wake up, as soon as I start to move around, I have to cough and get things moving. It's like my chest is really tight after sleeping. So, with that you can't help but think this is the CF in me that is doing this. 'Cause like a few years ago, that wasn't happening. Like when I was at school, I would wake up and maybe cough a little, but it wouldn't take long to get things moving. But now it's much more* ***there,*** *I suppose.*

Although people with CF have a chronic, life threatening illness, they do not experience themselves as being ill all the time. When they are well, they are less aware of their bodies. When they are *ill* they become very conscious of their bodies. As stated, health for them is a state of "unselfconscious being that illness shatters" (Baron, 1985, p. 609).

This chapter has discussed how persons becomes aware of the effects of CF on their health, they realise the personal meaning of the disease and are aware of CF in everyday life.

CHAPTER FOUR
LEARNING TO LIVE WITH CF

> *I suppose that they [Medical Specialists] expect me to be responsible for my own well being. It's up to me to take care of myself. I'm not living at home with my parents looking after me, I'm under my own care.*

This chapter discusses how people with CF learn to take responsibility for their own care. Their experiences are described by the themes of *learning to live* and *learning to manage*. While *learning to live* they are *learning limits* and learning to deal with the disease in the context of an adult life. They are also *making a conscious effort to look after self;* it does not always come naturally. *Learning to manage* includes *learning to manage illness, learning to manage hospitalisation* and *learning to manage relationships with health professionals.*

LEARNING TO LIVE

Learning, in relation to chronic illness, is often understood to be learning about the disease process or learning the routines or treatments associated with management of the disease. These form part of the learning about CF, as was discussed in the previous chapter and will be discussed later in this chapter. *Learning to live,* however, means gaining by experience the knowledge and skills to order or organise one's life. It describes the transition that people with CF make as they gain increased understanding of themselves and determine the adjustments they must make to look after themselves.

Learning limits

Although people with CF have had the disease all their lives, and have usually been involved in treatment regimes from an early age, further

learning comes as they start to manage their own lives. As they enter the adult world their lives broaden and they assume adult responsibilities; they must learn to know themselves and their limits. Two of the participants describe this as a process of self learning and said that, although they were given advice, they needed to work things out for themselves.

> *Sometimes I looked after myself and sometimes I didn't. You go through a funny learning curve. No one can tell you. It doesn't matter how much a professional tells you or how much a family member tells you, you still have to go through it yourself. You're the only one who can work out your limits, and then you have to work out whether you're going to listen to yourself. When you start working out your limits, that's when you start listening to people.*
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> . . . . . .
>
> *People can give you advice but the only way you learn is through your own experience and coming to your own conclusions.*

Adult life brings with it increased self responsibility. Adults with CF are now healthier than adults with CF were 10 years ago. They have greater choices, responsibilities and expectations than previously. All the participants are living active and involved lives. They are making choices about further education, work, and relationships. This was viewed as a good thing by one participant but he acknowledged that it also created extra stresses.

> *Now, if you are reasonably well through your teens, you're likely to go to university, or go to work. The weller you are, the more outgoing and social you are as well. You're going to meet a partner, get married or not get married, so those are all issues and they impact very much on your life. Working, there's a*

whole set of stresses that you don't really need. When you're at school you have to study but it's a safe environment. And once you start getting a bit older and whether you leave home or not, there's a whole another set of expectations on you. Those are stresses that affect your life.

One participant describes how she learnt to identify work stresses that she believes have negatively affected her health. It took her some time to develop this understanding. The responsibilities of adult life such as holding down a job and being financially independent had a higher priority than looking after herself at times. Now she is no longer working, she can see that the choices she made were not always the best for her health. So, she has now decreased the amount of stress in her life and increased her ability to look after herself.

Once you're working you owe a loyalty to someone else. You've got a job, you've got to keep it so you're not going to take a lot of time off work. Usually you're reasonably well but there will be times when maybe you should have stayed home and you won't have because you still have a loyalty there. [Work politics] add a stress to your life and its a stress you bring home. That's making you more susceptible to an infection because it's wearing you down when you don't need it. It's very hard to identify to start with too. They're an extra stress on your body and they tire you out. Those sort of things you need to take into account more.

I'm at home at the moment, I'm still busy doing but I can spend two hours on physio [physiotherapy] if I want at my own pace, nice and leisurely now. When I was working I got up, I did physio and if I needed a bit more that was just too bad, I had to go to work. So you're cutting out something that you really

needed, to go to work.. I would have my [nebuliser] 3 times only for the fact that I would have it immediately when I came home while being at home I could have it every four hours like they would give me in hospital. You can adjust your routine to be very good and look after yourself very well. I've got the time to do that whereas when you're at work these are things that get stopped and it's because you owe a loyalty somewhere else and you've got other priorities in your life. You've got a mortgage to pay, rent to pay, just normal bills to pay. And yes, you're doing your physio but its how quickly do you have to stop it, and when you come home and crash into bed because you're exhausted. As you get a bit older it's just life and life's responsibilities basically, and the fact that we never really had them before but there are a lot of us doing that now.

Other participants came to realise the negative effects of drinking alcohol, missing meals and lack of exercise on their bodies, especially as they got older. They learned to take better care as they realised the benefits this would have. Admi (1996, p. 177) also found in her study of adolescents and young adults with CF, that as they grew older "they began to acknowledge the difference that adherence made in their health, compared to non adherence". The participants learned that as they aged they had to take better care of their bodies. "Getting older" depended on when the person found their health significantly affected by CF.

I probably didn't realise how going out drinking all night, not eating for a few days on end... would affect me healthwise a few years ago. I'm more on to having to take care of myself, eating properly these days. I think that I probably could be a little more slack with my body than I am today. If I was doing the same thing I was doing three or four years ago now, I would be

sick a lot more of the time. So I think I'm more responsible now than I was in some ways.

. . . . . .

As you get a bit older life catches up with you - I know for me it has. And it wasn't that I didn't care...once your body gets a bit older and it loses its own bounce, you lose your elasticity so things catch up with you. And that's when it starts to change, you get old. It's not that you get old, old. It's just that your body changes and it doesn't bounce back the same way. And even if you had been really good to yourself, it's still got the changes, its got its own little clocking system and it waits. And it's no different. As you get older, you get more susceptible to things and it's the same with us. Sometimes it just happens a bit earlier. I think our body ages itself a bit quicker than yours. Well, if you have to fight infections really regular!

Between the ages of 15 and 25 you don't really give a damn about your body anyway so you're still dealing with the normal rebellion that you go through...we have a lot of stamina, that's what people don't always see, so you can often carry yourself through when you should have called it quits. Pay back time is usually after 25. If there's a time when you were a little less cautious than you should have been, you've let an infection go too far, there's scarring that you've caused or whatever, and just the fact that maybe you've run out of steam.

There was a recognition of the need to take better care because CF is accelerating the normal ageing process in this participant. The comparison of their chronic illness with premature ageing has been made by others (Register, 1987). Another participant who was relatively well did not have this experience.

It's not something that I feel necessarily happening to me at this stage. I haven't got a lot of other problems that people around about my age could have. By and large I feel pretty average that way. I feel as though if I hadn't have had this setback [CF], I would be stronger and healthier than I am now.

Making a conscious effort to look after self

All the participants describe having to make a conscious effort to be physically active and to look after themselves. The level of effort required depends on the individual and the severity of their disease.

We have to stand over ourselves and do it. And when you're a person who doesn't like routines, it doesn't really come very naturally.

Conscious effort is required to do active treatments such as physiotherapy, exercise and eating properly. A decision has to be made by the person to adjust their lifestyle to include activities that do not come naturally. One participant describes learning to take better care of himself. It was a conscious decision and required a concerted effort on his part.

You do have to keep better care of yourself and make sure you do. I just got a bit lazy, didn't look after myself properly. Didn't do enough exercise. I don't know why. But then I decided it was time to and now that I do, I'm glad for it. But you do need to make the effort.

I've just got a dog and I walk him a couple of times a day. I get a lot more exercise. I push bike more, I've started going to the gym. Just keeping a lot healthier. Putting on weight; trying to eat more. Last time I went [Medical Specialist] was really happy. Since the start of the year I've put on about 10 kg which

is a lot for me. I think it's time to try to keep out of hospital, I'm sick of feeling crook. I'm trying to make a difference. It seems to be working.

Another participant who was not so effected by CF is aware that his level of adjustment to the disease is not as great as some others. However, he has made conscious decisions to do activities that will be beneficial to his health.

I think I made quite a few little adjustments all around the place but I'm sure that most of the other people who have classic CF would say that people who have my level of severity, probably haven't had to make adjustments. And I certainly think about that too looking at people who are much less well than me. My level of adjustment is very minor. I still do a lot of the things I used to do before CF, and I want to. It's a conscious thing too, I don't like sort of giving up. In some ways I want to keep up the same level of activity but to change the focus of it so that certain things that are more beneficial to CF are accented. That's the way I consider it. Probably a more subtle approach to it.

The participants' conscious effort to look after themselves will be elaborated in the following two chapters, in relation to how much this effort effects living their lives. All the participants found regular physical exercise was beneficial to their health although it still required effort to do it.

I've been trying to make myself walk up and down the hill every day, and I haven't been doing it every day. But I've been doing it three or four times a week and I find that if I do that my breathing's much better.

The positive effects of physical exercise on their health is discussed in the next theme.

LEARNING TO MANAGE

Learning to live was not the only learning by the participants. For most of them, as they are getting older, they are experiencing more exacerbations of their lung disease. They are therefore also *learning to manage illness*, *learning to manage hospitalisation* and *learning to manage relationships with health professionals.*

Learning to manage illness

The participants were learning to recognise and manage exacerbations of their disease. Each person had a different pattern of symptoms and disease course. One participant describes learning that his disease was not necessarily going to follow the "normal" medical trajectory and realising that there are differences in severity of the disease that are not always clearly differentiated in scientific and medical literature.

> *Those were the days when I thought cystic fibrosis (CF) is cystic fibrosis (CF). There's just one disease for everybody and you're on this path and you'll go down at the same speed as everyone else. But I have since learnt after that, that is not the case at all. It comes in many guises really.*

The participants are learning to judge their symptom severity, and whether they need to be hospitalised for intravenous antibiotics and intensive physiotherapy, or whether oral antibiotics and treatment at home will be sufficient. Previous experience of an exacerbation allows them to make a better analysis of their situation.

Sometimes I'll get an infection and I can just get rid of it with oral antibiotics and I can be quite sick but just put myself on a course of ciproxin and a week later the infection's gone down. But then sometimes it's just resistant and so it's hard to tell. Usually, once it gets to the point where I'm having trouble just getting out of bed and walking round, that's when I know it's time to go down there [hospital]. You do leave it to the last possible minute but it's hard to tell because sometimes the pills will kill it, sometimes they don't. [Speaking about last hospitalisation] I could foresee it coming. Each time I catch it a bit earlier because I know what the progression is like.

. . . . . .

Sometimes you miss the first couple of triggers, and then I've got to go and see [Medical Specialist] and I'm working out in my mind whether I need to go in or not. Can we put it off another week? Can we just get some cipro [ciprofloxacin] - sometimes you can get a good dose of cipro and you're on your feet again.

Learning to manage hospitalisation

Most of the participants in this study had not experienced being hospitalised during childhood. As they grew older, some experienced a deterioration in their lung disease that required them to be admitted for intravenous antibiotics. Some also experienced gastrointestinal symptoms which required hospitalisation. They were unfamiliar with hospitals and had to learn how to manage this experience as adults. One of the difficulties of being in hospital is that it is perceived to be an institution for sick people who require treatment in bed. For the participants, the physically inactivity that is expected in hospital does not lead to an easier recovery when they get home. They find that it is better for them if they exercise as soon as they are able to.

I find it really hard coming home from hospital because you're not doing any proper exercise and you come home and you just feel lethargic. It's not that you're sick still, it's just that you've been sitting on a bed all this time and your muscles haven't drained away, but they haven't been active. I usually go up to the physio and use the exercycle so, when I come home, I don't feel like just crashing out. It's really hard because you don't have that in-between step, and if you've been in for two or three weeks, it's very hard because your whole body is out of whack and you don't have to do anything for yourself. You're just mooching around.

. . . . . .

I get better quickly when I come out. I just gotta make sure I stay on my treatment and exercise but I normally get better fast. When I come out I'm not 100%. I've got to work on it 'cause just lying in there, you lie around and get lazy. You come out and you've got to get back into exercising but all the physio in there helps makes you feel better. Normally, after about four days I start coming right.

One participant described how she had learnt to accept and tolerate hospitalisation and now lets the antibiotics run their course.

Last time I didn't push to go home as quickly. I wasn't feeling well still. But that's only age you see. That's me deciding well, maybe I'll let them [antibiotics] run their course now.

More about their approach to hospitalisation will be included in the following chapter in the theme *rejection of the patient role*.

Learning to manage relationships with health professionals

There were marked differences between health professionals who had an understanding of CF and acknowledged the person's own knowledge of the illness and those who had little or no understanding of the disease. One participant describes going to the Accident and Emergency Department, knowing he needed intravenous antibiotics for a chest infection, and having to wait for a doctor who is familiar with CF who could assess whether he was sick enough to be admitted.

> *Last time I went it was almost as if they didn't believe that I needed to be in there. And they called up a Registrar to come and check me out and she had to come out from her house. I'd already been poked and prodded and they said "We're not sure if you're sick enough" and I knew damn well that the only way I was going to get better was on IV [intravenous] drugs. And so I had to wait until this woman came along and gave them the go ahead. "Yes, this person really is sick. Give him a bed."*

The preference for treatment from health professionals who know and understand their disease is illustrated by an example where one of the participants had to be boarded in one ward because his usual ward was full. He was left waiting in a chair for several hours, perhaps because he was not obviously ill, and the nurse did not have an understanding of his condition.

> *I was glad to be there except the next day they found a ward for me but it wasn't Ward X. It wasn't where I should have been but it was the only place they had a bed. Then the next day this nurse she got me out of bed and she said "Right, you're being moved later on" and she changed all the linen and I ended up not having a bed for about four hours. She went off duty and the next lot came on and I was just sitting in this chair going "Oh, I want to lie down. I want to go to sleep". I think she was probably just doing what she was told to do and she didn't*

realise I was going to be in there a few hours more but she could have been more caring.

Another participant describes the lack of knowledge of CF that some doctors have. This is evident when they ask her how long she has had CF!

I still occasionally get asked how long I've had CF from a doctor. And that just infuriates me. I used to think well maybe they just slipped up and they mean when you were diagnosed but that's not what they meant.

The participants have a good rapport with their Medical Specialist and value this relationship because he appears to value their knowledge of their own illnesses; he is the health professional who really **knows** them and knows their disease individually. This relationship is seen by the participants as a partnership in the management of their disease. They realise the problem and judge when and how to present information to the Medical Specialist. But, this partnership is not shared or understood by junior doctors or nursing staff.

And they forget that we have a very good rapport with our Specialist. Most of us do. And if you don't to start with, you build one up because you're seeing each other regularly. And he knows when you need to go in, and he knows when you're going down but you don't quite need to go in, and you can probably between you work out what you're going to do. You compromise. But a lot of nurses and house surgeons aren't used to the fact that you have a very good relationship, or pretty open relationship, and you might know what you're talking about. And it's just the fact that we live with it. They might read about it and study it, but we're doing all the practical (laughs) - they just know the theory.

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I think that the [Medical Specialist] probably gives you more credit for knowing that you can judge your own level, whether you're sick or not. But a lot of the people who don't have much experience of CF, probably think you're malingering or whatever. I suppose from their point of view, you're not a medical expert but most people with CF are.

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I normally tell him. I ring him up and say I'm not feeling well. I go and see him.

The Medical Specialist has gained his experience over several years and has treated many different people with CF. The Medical Specialist is an expert in disease management. Other health professionals are perceived not to have this understanding. Some of the participants would also like what they describe as a more "holistic" approach to their illness management.

When you go to your doctor and you're reasonably OK you're not going to tell him that there's a few office spats because it's not important. But those are the things they need to start taking into account more...They're an extra stress on your body and they tire you out. Maybe they need to monitor your vitamin levels or things like that at certain times in your life. I think they need to keep a close eye on what's happening. And take over from you a little bit. It's teaching us to be a more aware of our bodies as well as the doctors and what's going on - it's like this holistic approach - it's not really about what you present in front of them with - it's what's going on in your whole life and where you are.

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He asks me what's happening, what I'm doing but not really in any great detail. Like, give me a rough overview of your life.

This chapter has discussed how people learn to live with CF and learn to manage their illness. These observations have implications for the healthcare of people with CF which are discussed in Chapter Seven. The next chapter will discuss how the participants are getting on with their lives rather than focusing on their illness.

CHAPTER FIVE
GETTING ON WITH LIFE

> *You don't say very much usually. And because of that, people don't think that you're very sick so that works against us as well. And the fact that you usually try and be independent yourself, so you're working against yourself all the time. But all it is, is that you're just getting on with life, that's all it is.*

This chapter discusses the importance the participants place on *getting on with life* rather than focusing on their illness. Getting on with life means living a life which is not overwhelmed by CF. They value the time in their lives when they are not aware of their illness; when they are only aware of all the other concerns that make up their lives; the state of unselfconscious being. *Getting on with life* includes *balancing treatment regimes, minimising illness to self and to others*, and *rejecting the patient role.*

Balancing treatment regimes

The participants' desire to get on with life means that they make choices about how much time and attention to give to treatment regimes. They may choose to risk a decreased level of functioning by deciding not to follow through with treatments which seem to prevent them from getting on with life.

The participants did not follow advice or regimes to the letter if they believed this would impinge too much on their life. They did chest physiotherapy when they thought they needed it rather than according to a daily regime. One participant describes how, when he feels well, he doesn't always do his physiotherapy, but when he is unwell he arranges for a district nurse to help him with it.

I don't do it every day. I only really do it in the morning 'cause that's my worst time. The rest of the day I run around and that, so it clears out itself. Yeah, I always do it in the morning 'cause I know if I don't do it in the morning then I'll have a crappy morning. But when I'm real bad, I know. I sometimes try to stay out of hospital and so then I'll get the District Nurse to come round. It's just something that has to be done. She comes round a couple of times a week and does the physio. Only when I'm bad it helps sometimes.

Another participant describes how she only does physiotherapy when she is unwell. When she is well she does not do physiotherapy because she does not *need* to. She can get away without doing it. So, although she knows it probably would be better for her, it does not fit into her life.

I don't actually do any [physiotherapy] myself except when I feel I'm getting ill. That's usually shortly before I go into hospital so it's kind of shutting the gate after the horse has bolted...I do everything that I have to do without doing the physio. So it's like I don't feel I need to do it to get on with my life. Everyone tells me it would be beneficial if I did it and I'm sure it would be. I'd probably be fitter than I am. But I just don't do it. I just lead a sort of unstructured lifestyle so I don't have a set timetable of things to do. So I don't have a physio time slot in my day (laughs)...when I get an infection my chest becomes so clogged up that it's difficult just to walk around the house. So I have to sort of clear it out just to be able to breathe easily. So I do it as a last ditch kind of thing.

Part of the reason a treatment regime may not be adhered to is that it does not come naturally or fit easily into their everyday life. The person has to

expend mental effort to complete the regime and this conscious attention detracts from getting on with life.

One participant explained how someone who has a disease may not necessarily comply with treatment regimes, even if he knows they will be good for him, if he cannot fit them into his working life. He spoke about a new drug that had to be taken four times a day.

> *The thing that they forget is that they can say this is a wonderful technique but if it takes too much of your day up, and you're a working person, then it's not a good technique. You want something you can do with ease. A lot of people pulled out of trials. Not if you're working. Get real, it inflicts on your life. And most people say "So if it's gonna be better for you, you just do it don't you?" But you still are trying to keep a reasonably normal life, well not normal but an active life so that you can feel independent, and those things just intrude too much.*

These participants' stories clearly indicate that adults with CF decide what level of care to give themselves, and risk the effects that this may have on their health and level of functioning.

The need to get on with life or have some parts of his life continuing normally was particularly strong for one participant. He describes how he attends social occasions at physical cost to himself.

> *Because you can do your own tricks, you go out and you're fine, then you come home and you crash, you literally let your air out, and I'd be really tired. Why do I do this? I have to go out to keep sane, I can't go out there and tell them how I'm feeling, like I'm really grotty. I want to go out and still enjoy. It may be*

> *that you don't go out and dance all the time, but you go out. You're at the table, you still go to dinner. You might not go as* ***late*** *but you're there for the evening. And you'll be fine, you'll be a social person while you're there. And you come home and you think "Oh God" which is what people do anyway.*

Another participant describes how he tolerates hospitalisation better if he can go home for part of the day so that his entire life is not taken up by his CF.

> *I have a compromise too, if I can get out in my afternoons between doses and have a good few hours where I feel sane, then I'll stay in for his [Medical Specialist] extra couple of days, I don't mind that. I need to have some of my life continuing on normally, I need to have that.*

This apparent rejection of the patient role will be discussed in the next section of this chapter. Admi's (1996) study of adolescents and young adults with CF similarly found "the informants repeatedly emphasised that the disease did not play a central role in their lives and they made continuous efforts to prevent CF from interfering with their daily and future plans" (p. 172).

Minimising illness to self and to others

The value that the participants place on *getting on with life* means that they generally do not dwell on their illness. This was seen as pointless; they had the disease and they had to get on with it.

> *Just got used to having it. Go along with it. Not much else you can do; you can't dwell on it or anything. You can't really change it. You've got it, so you might as well...I just know that I've got it so I just don't think about it very much.*

> *We're not a lot different than anyone with cancer or AIDS but we're not taken in the same light. And most of it is our own fault... because we don't dwell on it. We just get on with it and do it.*

Not wanting to be so aware of their illness means that some people at some times choose not to be involved with the Cystic Fibrosis (CF) Association.

> *You go through a stage when you're quite well and coping on your own or you're not well, but either way you don't want to have anything to do with CF. No one else is doing you any good. And if you're really well and off doing your own thing then you don't want to be reminded that you're likely to not be well again or you're so busy living your own life, you haven't got time for it. So you've mostly got people involved when they're middling to not well or unless they're just going to be involved anyway or just not well. It's just a support group, they come and go when they need it. In their late teens you don't want to know usually, most of them don't.*

Another way participants are *getting on with life* is by not talking about their illness with others. By not acknowledging their illness to others, they are less conscious of it themselves. One participant described how she usually did not tell friends the details of how she was feeling physically as this stopped her from feeling so ill. This meant that they did not always perceive how sick she really was.

> *I think that's also part of your own defence mechanism too. If you don't have to say "I'm not well, I'm sick" all the time, then you don't always feel it either. So you don't want to affirm all the time that you're feeling as grotty as you feel. Well, I know I*

don't, so in that respect it's easier to say "Oh I'm OK" or "Not bad". Those things just cover multitudes...You don't say very much usually. And because of that, people don't think that you're very sick so that works against us as well. And the fact that you usually try and be independent yourself, so you're working against yourself all the time. But all it is, is that you're just getting on with life, that's all it is.

There are also choices to be made with social acquaintances. To be perceived as someone with a chronic illness opens one to misunderstanding, pity and being seen as less of a person.

It's like when somebody asks you what's wrong with you, you have to weigh up who they are, whether they need to know and if they need to know, what they need to know. If you start going on exactly what CF was you'd freak everybody out. Then they treat you so nicely and kindly. Oh God... It is difficult but you don't want to be labelled with a sick label, OK. So, often I say that I've got asthma. If you're gonna deal with someone on an ongoing basis then you've still got to weigh up whether you should or you shouldn't. Sometimes you don't want them to think of you as anything else than the person you are. Or sometimes you need them to have a little bit of understanding that you might not feel wonderful at times.

The nature of CF means that the signs of the disease can be concealed or passed off as a different illness such as asthma which requires less explanation.

CF's really insidious, it's really nasty. I mean, unless you're really ill, you don't always see the effects. You can easily mask it apart from your breathing, but people will think that you've

got asthma so that's easy, without going into a full description, it's easier to say yes.

Admi (1995), when exploring the management of disease related information among adolescents and young adults with CF, found that they chose to tell people who

> ...had the ability to express caring, deal with the information and keep symmetric reciprocal relationships...The informants with CF reported dissatisfaction with being treated like patients in social relations when it was not relevant. People with CF...seek mutuality within relationships in which their sick label is known and do not readily accept the transfer of their sick role into other social roles when they perceive the sick label as irrelevant" (p. 491-492).

Rejecting the patient role

Focusing on getting on with life leads to a minimisation of illness that extends to having a different concept of being sick from people who do not have a chronic illness. "Being sick" was variously described by the participants as "feeling awful", "feeling horrible", "having trouble getting out of bed or walking around", "difficulty breathing" and "no interest in getting out of bed". They live their own pattern of periods of illness alternating with periods of relative wellness.

We go up and down all the time. I mean you can be feeling really ill one day and all right, not wonderful but enough to cope, the next.

The combination of being used to managing their own illness, wanting to get on with life, needing to be physically active and having a different definition of being sick, leads to a rejection of aspects of the patient role in hospital.

The person with CF may not perceive themselves as being ill although they have a disease and are experiencing signs and symptoms and are perceived as being sick by others (including health professionals). This difference in perception may lead to conflict with the expected patient role.

> *We used to be put on bed rest when we first came into hospital. I can remember visiting [friend] one time and she was just livid. Oh boy! And it was because she had a new house surgeon and the state of her chest at the time, he decided bed rest is what she needed...Most of us don't really want to be tucked into bed and given a hot water bottle and read a story. And if you go and visit other people in other hospitals, it's the same. Most of us get dressed unless you're feeling rotten, you just can't be bothered and that's when you need to be in... all it is is that we get sick as a normal course of our life so we don't treat it any different. I remember a social worker coming to see me many years ago and she wanted to know if I was alright about being in hospital and whether I understood why I was there. Did I understand the treatments I was having and why I was having them - you just don't need it! We usually know why we're there - we're sick. A lot of people who go in don't understand why they are there and they don't understand their treatments, but most of us do.*

One participant describes the conflicts he had coming to a new ward with staff who did not know or understand adults with CF. He was perceived by them as being sick and requiring a wheelchair to go to the X-ray department; he did not perceive himself as being ill because he could still walk.

> *And that was something I found really hard coming to that ward because they didn't want me to get up and move around and I used to have big battles. I think [nurse] used to get cross*

with me. I don't like going to X-ray or breathing tests in the wheelchair. And a lot of it's me. I just don't like the fact that I'm in a wheelchair. I hate it. If I need it I'll make use but if I can walk I'd rather walk. I don't care if it takes me half an hour and they could have got me there in five minutes, it's what I prefer and there's no reason why I shouldn't do it that way. And they found that very hard, I wouldn't just conform into a wheelchair and get wheeled around somewhere and brought back. But I wasn't ill - I could still walk. And I was walking within the hospital and it wasn't cold or draughty or anything.

They tend to minimise acute exacerbations caused by chest infections and this in turn leads to their regarding hospitalisation in a different way from other patients and the staff.

It's just an infection, or it's an infection and another infection, or whatever. Or just a tune up - that's what I call most of mine. When you go in and get cleared out and come home again. And that's just usually over a week or so and that's all they are. But you don't need to be treated extra special...Those are the things that are particular to ***us*** *and the type of illness we have and although we're sick we're not usually out and out just victims or patients that need to be just put in bed.*

Hospitalisation for intravenous antibiotics is accepted as necessary when they are sick but some find this difficult to tolerate once they start feeling well again.

I don't like going into hospital but I like having a massive infection even less so, by the time it gets to the point where you do have to go into hospital, it seems like it's going to be quite

nice to be in there and get that drip. But then two weeks later it's like "Let me out, let me out!". Going stir crazy!

. . . . . .

Normally when I first go in I don't mind because I just feel awful anyway. But having all the drips and that annoys me. Sort of hurts and that. After a few days, after a few days of that I just get sick of it. I want to go home again but to start with I normally don't mind it because I'm so...because I don't feel well and I know that its doing me good. I don't normally stay in there long. A week or not much over a week.

. . . . . .

And usually that's when you need your rest when you can't do it [intensive physiotherapy], when you're not interested in anything. You're not interested in even getting out of bed and making yourself a cup of tea. And that's when you need to be there. After that, when you're starting to feel a bit better, you just need to get your system back to normal. I find it really hard coming home from hospital because you're not doing any proper exercise and you come home and you just feel lethargic and it's not that you're sick still. It's just that you've been sitting on a bed all this time and your muscles sort of haven't drained away but they haven't been doing anything. They haven't been active.

The inactivity of patients, expected by health professionals in hospital, was particularly difficult to deal with and was not considered by participants to be good for their physical health. But compromise is possible.

So we've come to a few compromises, we've worked out things that suit us both. That's age as well. I would have never done that before. I would have, but I would have begrudged doing that. But I can see now that it's more appropriate to do it now

*and **also** I don't feel so drained when I come home as well. You let it work its little pace.*

This chapter has discussed how the participants do not dwell on their illnesses but strive to get on with life. Balancing treatment regimes, minimising illness to self and others and rejecting the patient role are all ways they find to reduce the impact of illness on their lives.

CHAPTER SIX
FIGHTING TO GET ON WITH LIFE

> *There's this aberration you can feel, like you're fighting something which is essentially you and then other times you can put it in a little box and say "It's a part of me but there so much more to me as well."*

This chapter will discuss how when the participants experience their health as deteriorating or unstable, they work harder at maintaining their well-being and stopping the disease intruding on every aspect of their lives. They experience themselves *fighting against CF* in order to get on with life, *redefining what is important, envisaging a future* and *fighting to maintain a positive attitude.*

Fighting against CF

When people with CF are unwell or experiencing their health progressively deteriorating, getting on with life, although more difficult, is still important. In unstable phases, people experience themselves fighting the disease, fighting their own bodies and fighting themselves. One participant talks about the genetic nature of the disease, that CF is in every cell of his body. When he is fighting the disease he is really fighting himself. At other times, when he is less ill, he can separate the disease to just being a part of himself.

> *I think in depressed states, that's how you can think of it, that you are actually fighting yourself. Especially seeing it's genetic and you think about seeing it in every cell of your body. There's this aberration you can feel, like you're fighting something which is essentially you, and then other times you can put it in a little box and say "It's a part of me but there so much more to*

me as well." Positive attitude, keeping it in perspective is important.

Another participant can imagine what it is like for others although he is not experiencing a deterioration in his own health. He thinks about the disease affecting his body and understands the experience of others who, when experiencing bodily changes, may feel that that the disease is taking them over. The times when he has lost confidence in his body he has had a similar experience.

I think every now and then I 'spose you look at your body and you think "Is your body changing and conforming with the stereotypes of the disease, the very skinny people, the protruding bones, the changes to the fingers and so on? That finger's a bit bigger than it should be and this knuckle looks a bit wider and slight loss of weight and so on. Is this the beginning of a much faster decline?" But I think those were the days when I thought cystic fibrosis (CF) is cystic fibrosis (CF); there's just one disease for everybody and you're on this path and you'll go down at the same speed as everyone else. But I have since learned after that that is not the case at all. It comes in many guises really. I look for those signs all right and I can quite understand that people, particularly maybe people who spend a lot of time with other people who have CF or feel maybe they might be a bit isolated because of their CF, I'm sure they feel as though there's a thing which is taking them over and turning them into a certain type of person who looks a certain way. It's almost like being a dwarf, or whatever. I don't feel that way, it just hasn't got that far down the track. Some people I've heard describe it as being like a friend, it's like a familiar...I don't feel that way about cystic fibrosis (CF) at all. Certainly I don't feel as though its like a menacing shadow that's sort of stalking you all day ready

to ...when I talked about a loss of confidence there's that sort of feeling as though you're being stalked and suddenly you've just been gripped by this disease which is there, ready to take hold if you go a little bit out of balance.

When the body is more greatly affected, the ability to get on with life cannot be taken for granted. As the disease becomes more progressive, CF is perceived as having an increasingly bigger impact. The disease becomes a constant presence. One participant describes never being able to take a holiday from CF.

And that's where CF dictates with [partner] and I as well, when I'm not very well. Then your time as a couple is gone. It's eroded and that's what people in the health system do not see. They think it all goes on dollar figures but they do not see how much of our lives are spent tied up with CF. What saving you do do... if you need a holiday, you need a holiday. If you're spending all your money on medication and hospital bills, you don't get a holiday for you as a couple. You're not doing anything good for yourselves. You need time out. A parent can actually take time out, they have social welfare. So you can nominate another caregiver 28 days of the year. We can't. What do I do? Do I pack it in a wee box and leave it at home under the bed? I have to take it with me, so in that respect we don't ever get a holiday.

The same participant spoke about times when he is fighting the disease for control of his life. The disease has control and he has to find ways round it. He has to fight to maintain his independence.

It is really frustrating because it does take a lot of your life. I don't think about it like that very often, because you just get on

and you do it but it rules. We were at a meeting the other night and I said "It's like a split personality - it decides whether you're really going to do something or not - you are a placator. You try and work out how you're gonna get round it. You don't have the control". And if you're very well, you usually can have much more of a control but it just depends where you are at the time. And that's a chronic illness syndrome basically. Most of us try and be very independent people and you're fighting against yourself basically. But that's just how it is...You have to work out how far do you let the intrusions in. When do you surrender? People think that's the wrong way of looking at it - that you're surrendering - but inside, that's how you basically feel. These are the battle lines. It is a lot of compromise. Because we don't lie down and be pathetic patients. We never do it only as they said or when they said it. I don't think we would have survived if we were that sort of people. We'd still be on bedrest (laughs).

Redefining what is important

To cope with changes in their health and their reducing activity level, the person must redefine what is important to them. If a person has to give up work, the focus on what is important in his or her life must change. One participant who fought very hard to live as normal a life as possible and valued his ability to hold down a full time job, speaks of how he now values different things in his life.

I think it's lucky really, 'cause in many ways a lot of people don't expect us to carry on working or to work full stop, so we have a bit more freedom than everyone else. And yet that's what I always used to work against, was the expectation that I probably wouldn't, 'cause that used to annoy me. Why would I probably not [work] when I'm quite capable of doing that? And

now I'm quite happy to say "Well OK". But I'm gonna do something out of it which is the difference. It does change and it depends on how your health has been through your life. Most of us have real ups and downs, some of us cruise. It all depends on how far you work, but generally you retire earlier than everybody else. And I don't think that I can see a problem with that, I think that it's good.

Another participant who was working and experiencing a very stable phase also talked about being prepared to redefine what was important in his life when his health deteriorated.

I 'spose the thought that I might one day not be able to work, it is in the back of my mind. But I think then there'll be quite a few other important things that will take over precedence by then. If I can't work there'll be quite a few other worse things happening, I think. So I'm not letting myself get too fazed out by that. That's probably an area where I'm very conscious of having to think of the next phase well before it happens really. I certainly want to be a couple of steps ahead of the game. I don't want to be in some sort of a slightly debilitated state and yet feeling incredibly frustrated because I'm not a worthwhile member of society or something. That would be doubly stressful. So I'm certainly very conscious of having to put things together in a way which means that I can work at a lower level and get other satisfactions and put the extra time I might have into better management of finances or better management of my own health. And if there's no financial recognition of the things I do, well I'm quite happy to do things in the community or something like that. I haven't got to that stage where I've worked out what the right mix of things is.

Envisaging a future

Envisaging is imagining your future situation and how you might cope with it. The participants see knowledge as an important tool in helping them cope with and prepare for future phases of their disease. Knowledge came informally through sharing the experience of others. Discussions with other adults with CF were seen as valuable. One participant talked about lung transplants and how he had discussed this possibility with other adults.

> *I've never had a conversation with [Medical Specialist]. We've talked about transplants like we've been talking about them, what's happening out there and passing information on, but on a personal level, affecting me, no never. And it won't really happen 'cause those are the extremities of treatment and until it's really necessary you don't discuss it. But* ***we*** *discuss them, discussed within the group - we need to - what you're basically doing is basically getting yourself ready, in case anything comes up. It's not a shock, it's not such a surprise. You don't have to handle a whole lot of things. You've talked about it. You're still talking about it in the third person, so it's not affecting you. But you've still gone through the motions and that's why we discuss a lot of things and that's why the newsletters are there. We need to do that. You need to be able to identify the thing that you're dealing with, even if you're not dealing with it at the time, even if there's no hope at the end of it. But the fact that you need to identify it, to identify how you're going to get round it, how* ***you're*** *going to cope with it, and that's the way we deal with it.*

They wanted to face up to the reality of what was happening with their disease and wanted this information from doctors. This helped them prepare for the future and cope with the present.

I think that's why we have problems with some of the doctors because they're not used to patients dealing with things like that. We don't want a lot of bullshit - we want to know up front what's happening and so what if it's the worst of the worst news they can tell us. You need to find the way round to deal with it. So that's the difference, I think with a lot of us, is that every step along your life things have changed, the disease has always changed. Sometimes you get a sort of a, not a remission but a reprieve, and you go off and do your own thing for a while but there's always a payback. So you've always just had to deal with it all the time. So you're quite used to doing that and it's very infuriating and frustrating if you're not allowed to do that. Because then what they're doing is that's part of our coping mechanisms and in denying that it means we can't cope. Not that we can't cope but what do we do next?

Those participants who were well were either conscious of their future possibilities and did not try to plan too far ahead or they chose not to think about the future at all.

I seem to have something every few years but there's been nothing that's shaken my confidence to a great degree for quite a while now. For the last four or five years I've been in a pretty good phase really. I take it...don't try to look ahead too much...I've been associated with other CFers who I've seen deteriorate very quickly. I don't think that's going to happen to me but it could do. I think in some ways I'm more likely to be run over by a bus than that to happen. I'm not really a fatalistic person but I don't rule out that there could be nasty unexpected surprises around the corner in people's lives. So for that reason I don't like to set up weighty schemes that are

reliant on being 100% well for ten years. I think a lot of people with CF, I've heard it said, that they just live for the moment or they just want to enjoy the next year or whatever it is. I don't think my horizons are that short, well they're quite a bit longer than that. But I certainly am aware that...I don't like to postpone.

. . . . . .

I just take it as it goes, I don't really think about the future much or ever.

Fighting to maintain a positive attitude

When people become more conscious of the "disease" and the effects it is having on their bodies, this in turn can affect their psychological well-being. One participant has to work hard to maintain his psychological well being whereas others who were not experiencing a deterioration in their health do not. He talks about the conscious effort he has to make to exercise every day and the benefits of physical activity on his psychological well-being.

I've been trying to make myself do daily exercise, walk up and down the hill every day. And I haven't been doing it every day, but I've been doing it three or four times a week. And I find that if I do that, I'm much more, my breathing's much better than if I'm sitting around all day doing nothing. And that makes you better psychologically, if you're more active.

Exercise has an important impact on psychological well-being for people with CF. Inactivity is believed by the participants to be bad for themselves, both physically and psychologically. Other strategies a person may use are positive affirmations and social support.

Keep a positive attitude. Affirmations. Seeing friends helps a lot. If I'm having a good time socially, I'm usually in a better

mental state than if I'm sitting home alone doing nothing, although I do like my times of solitude.

People have to put considerable effort into looking after themselves physically and maintaining their psychological well-being. By doing this they maintain some sense of control over their lives rather than just giving in to the disease or their "fate".

I think it all depends on the decisions I make. If I can keep my depression at bay and eat properly and exercise regularly then I can achieve anything. But on the other hand, I can see myself just frittering away my existence and getting sicker and sicker and wallowing in it. Letting the disease get me instead of fighting it...It's a progressive thing. It slowly gets worse. I've seen my chest x-rays and they're more cloudy than they were a few years ago. There's only one way it's going to go, providing they don't come up with a miracle cure tomorrow. It's going to get worse until it kills me. But I think I have control over how long it's going to take. If I just let it get me, it could be a couple of years, but if I fight it, it could be much longer...It seems like a really threatening monster sometimes and whatever I do there's no escape from the monster, its going to get me in the end. But I try and keep up a positive attitude.

For those who are unwell, death is tangible but they cannot live as if death is pre-eminent; they push it away, they cannot live that way. One participant describes knowing that if he gets a bad chest infection he could only have a matter of weeks to live but he cannot live with being so conscious of his death.

I don't believe that I live as if I'm going to be dead within a year. I just keep living and doing my own thing. I think it

would be impossible to live that way and I think if you believe something like that strongly then it will happen. So I just push it away. Not to deny that I have the disease but not to have it right there in front of me most of the time.

CF makes you more aware of your own mortality. I think having CF has caused me to be come a much more spiritual person than I would be if I didn't have the disease. Just for the fact that [I] know my own death - I'm much more aware of it than most other people - it a much more tangible thing to me. I could catch some antibiotic resistant Pseudomonas and go downhill in just a matter of weeks.

There is a strong sense of inevitability and reality in the way that three of the participants spoke about their own deaths. But it is something they do not dwell on. This chapter has discussed the strategies and increasing effort people with CF makes to get on with life as their health deteriorates. They experience themselves fighting against CF.

CHAPTER SEVEN
DISCUSSION

> As nurses we are in the privileged position of being able to share with young people who have a wisdom not born of old age, but who live life to the full (Duncan-Skingle, Hitchcock & MacDonald, 1992, p. 14).

This chapter will discuss and interpret the findings of this study in relation to the existing knowledge of the experience of living with CF and other chronic illnesses. The implications of the findings for people with CF, nurses and other health professionals, and people with chronic illnesses will be considered. The limitations of the study and the direction of future research will be discussed.

DISCUSSION AND IMPLICATIONS OF THE FINDINGS

This study reveals the experience of adults with CF to be one of getting on with life rather than dwelling on illness. "Contrary to an image of chronic ill health and disability, a high proportion of adults with cystic fibrosis (CF) are living full and productive lives" (Walters et al., 1993, p. 549). The participants in this study are positive people, living their lives with meaning and purpose. The four major themes identified in this study are:
Realising CF
Learning to live with CF
Getting on with life
Fighting to live with CF

Realising CF

Realising CF means gradually conceiving their disease as real and understanding the implications of the disease. Realising is also being aware of having CF in everyday life. The subthemes are:

Realising the meaning of CF

Realising CF in everyday life

The experience of adults with CF is shaped by their having had the disease all their lives and by the embodied nature of being a person. *Realising CF* reveals that there is a difference in experience between those people with CF who know that they have the disease all their lives and people who are diagnosed with a serious life limiting disease later in life.

People who know that they have the disease all their lives do not experience a period of disruption caused by the onset of symptoms and then the quest for diagnosis which almost always accompanies chronic illness (Corbin & Strauss, 1988). They have the symptoms and knowledge of their diagnosis all their lives.[2] For the person with CF, "disease can exist in the absence of illness" (Dimond & Jones, 1983, p. 5); and thus the person can have the disease without perceiving themselves to be ill. The participants are at times aware of having disease without being aware of being ill. Changes in their bodies alter their perceptions. Although they have had symptoms all their lives, these are taken for granted until significant changes in the body and changes to their embodied selves cause them to *realise the personal meaning of their CF*. They also needed enough maturity to be able to understand the medical knowledge of the disease and its consequences.

For those who have been aware of having CF all their lives, this life limiting disease is not so shocking as the experience of having other diseases that may limit the lives of young people such as cancer and AIDS.

> *We're not a lot different than anyone with cancer or AIDS but we're not taken in the same light. And most of it is our own*

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[2] The experience of parents when a child is diagnosed with cystic fibrosis is outside the scope of this study but has been described by Whyte (1992) as an assault to self image and an experience of suffering caused by the life threatening nature of the disease.

fault... because we don't dwell on it. We just get on with it and do it.

The phase of learning to accommodate a chronic illness described by Corbin and Strauss (1988) is also experienced differently because the disease has usually been part of their identity and life story since they can remember. Conscious "accommodating" occurs when they begin *realising the personal meaning of CF,* usually some years after they have been diagnosed. This aspect of accommodating the illness may be very gradual as there is a growing realisation of CF.

In this study, the participants whose CF was not diagnosed when they were infants had a different experience from those who were. Their experience was more like the descriptions of others who have been diagnosed with a chronic illness after childhood. The diagnostic phase was significant and the accommodating phase was not so gradual or delayed.

An understanding of the process of *realising CF* may assist families and health professionals to understand that healthy young adults with CF who do not appear to accept their projected lifespan limitations cannot conceive them as real when they have not yet experienced any significant change in their taken-for-granted bodies. Also, if the person has not experienced a deterioration in their health or *realised the personal meaning of CF,* envisaging the consequences of progression of the disease may be inappropriate for them. Minimisation and, at times, denial of illness should be seen as appropriate responses in people who want to emphasise getting on with life rather than allowing the illness to overwhelm their daily lives.

Learning to live with CF

Learning to live with CF involves the participants learning their own limits, learning how to manage their lifestyle in order to maintain their health, and

learning to manage treatments and their relationships with health professionals as they learn to look after the disease. The subthemes are:

Learning to live

Learning limits

Making a conscious effort to look after self

Learning to manage

Learning to mange illness

Learning to manage hospitalisation

Learning to manage relationships with health professionals

Although the participants had the disease during childhood, *learning to live with CF* largely took place during adolescence and young adulthood. It occurred concurrently with learning self responsibility in relation to other parts of their lives. They identified that much of this learning was no different from a person who does not have a chronic illness; they learned their own limits and to look after themselves. The different learning they did was associated with managing the illness, hospitalisation and relationships with health professionals. They all stressed the importance of self learning.

There is little mention in the literature about how young adults learn to live with a chronic illness. Strauss and Glaser (1975) and Corbin and Strauss (1988) discuss the strategies required to manage chronic illness by the person and family but they do not illuminate how this is learned. There is similarity between their descriptions and the effort required by the participants to manage their illnesses. For these participants, being told by a health professional was not learning; learning experientially, "for myself", was how they learned.

Admi's (1996) study of the life history of adolescents and adults with CF also found that "as individuals with CF grew into adolescence, they developed a better understanding of the disease and its implications and, at the same time, realised that the treatments had a positive impact on their

health condition" (p. 180). In her study, research participants identified two main areas of work: managing information and managing regimens.

The findings of this study are similar to those of Strauss and Glaser (1975). Most illness management is undertaken by the person with CF in their own home rather than by the health professional in an institutional setting. The study reveals the disruption that hospitalisation is to the participants' lives and also indicates that the lack of activity expected of them was detrimental to their recovery. The study also reveals the lack of understanding of the disease and the experience of chronic illness by some nurses and doctors. The participants wanted a partnership with health professionals where their own knowledge of their disease was respected.

The finding that hospitalisation is perceived as disruptive to their lives, and that they feel that the inactivity expected as a patient hinders their recovery, supports the argument that has been made for a shift of resources in the health care system to home care (Glaser & Strauss, 1975; Strauss, 1987). When hospitalisation is unavoidable, people with CF are better with health professionals who have some understanding of their disease and individual experience. Continuity of care by health professionals who have specialised knowledge and experience from caring for people with CF is the ideal. The present employment patterns and structure of most nursing work does not allow for the continuity of care that is required to provide holistic individualised care. Nursing positions that span hospital and community settings would better facilitate nurses' ability to assist people with CF to live their lives rather than to simply look after their disease. Adults with CF ask to be partners in their treatment and to have their own knowledge of the disease acknowledged by health professionals. They are the "central agents of management" (Strauss, 1987, p. 33) and deserve full recognition of, and support for, this.

The importance of experiential learning suggests that health professionals and parents need to encourage self awareness and self responsibility, and support adolescents and young adults while allowing them to work things out for themselves. Teaching is not simply passing on information or giving advice. Benner (1984) has described competencies of the "teaching - coaching function" in expert nursing including: "timing: capturing a patient's readiness to learn", "assisting patients to integrate the implications of illness and recovery into their lifestyles" and "eliciting and understanding the patient's interpretation of his or her illness" (p. 79). Nurses with this expertise and understanding of CF are employed in some CF treatment centres but often with a predominantly paediatric focus (Glew, 1993; Moore, 1988).

Getting on with life

Getting on with life involves the participants making the normal activities of life a priority rather than making the demands of the disease central to their lives. The subthemes are:

Balancing treatment regimes

Minimising illness to self and to others

Rejecting the patient role

These adults with CF emphasise *getting on with life* and not dwelling on illness. This process of getting on with life leads them to make decisions about whether to modify treatments if they interfere with the rest of life. Admi (1996) also found "later adolescence was characterised by finding individual strategies of routinizing and modifying the regimens in ways that ensured the maintenance of their health status and minimal interference with social life"(p. 180). If the focus of care is only on the disease and the need to prevent it from worsening, then they are not seeking what health professionals would consider to be the best possible or most achievable health status. But, by considering alternative ways of looking at health, they are looking after their disease as best they can without letting it take up too much of their lives.

Lieb (1976), in a discussion on the value of health, concludes that "health, then, of itself has no large or notable value. The standard by which it is measured...is the fullness of life; and it is that, of course, with which we have to be most concerned" (p. 169). McWilliam, Stewart, Brown, Desai and Coderre (1996, p. 5) found that, for older people with chronic illness, "...health simply means being able to do the things they want to do" and that they identified health as something different from their chronic illness. It was "...a sense of fulfilment or purpose in life through doing".

Roberson (1993) in a study of patients' perspectives on compliance found

> ...patients define compliance in terms of apparent "good health" and seek treatment approaches which are manageable, liveable, and in their view effective. Thus they develop systems of self management which are suited to their lifestyles, belief patterns and personal priorities. While many of these people would be labelled as noncompliant by health professionals, they see themselves as "doing a pretty good job" (p. 7).

Hunt et al. (1989) found that patients strive to fit treatments into their everyday routines in ways that will allow them to feel better while not making drastic changes to their lives. These observations are strongly supported by the experience of the participants in this study.

Health within illness is a concept discussed by Lindsey (1996). She suggests that by continuing to focus "...predominantly on the illness experience...a large part of the person's whole experience is missing and therefore denied. Because people experience their bodies as a whole (Merleau-Ponty, 1962), the focus needs to shift and equal attention given to the health experience of chronic/disabling conditions" (p. 466).

Minimisation, denial and avoidance are coping strategies people use in response to stress that have been identified by psychology (Folkman & Lazarus, 1980). Minimisation and denial have been found to be used by adolescents and adults with CF (Strauss & Wellisch, 1981) and their parents (Whyte, 1992). *Minimising illness to self and others* did not mean that the participants denied that they had CF or the severity or seriousness of the disease. Simply it allowed them to get on with life and not to be overwhelmed by their own or others' feelings and perceptions about the illness. "Once the illness becomes social, the social interactions of the ill person and those around him are modified. He is treated differently because he is sick, and he responds to others differently because he is considered sick" (Dimond & Jones, 1983, p. 6-7). As one participant said about not wanting to think that he might be dead within a year:

> *I think it would be impossible to live that way and I think if you believe something like that strongly then it will happen. So I just push it away. Not to deny that I have the disease but not to have it right there in front of me most of the time.*

Register (1987), from the perspective of a person with a chronic illness, writes:

> We learn to accept, or acknowledge, the seriousness and permanence of our illnesses, and then practice a selective denial. We can live as well as possible only if we ease up on the pervasive consciousness of illness to make room for all the other thoughts and feelings that a full life requires (p. 197).

The participants could be described as having the hardiness characteristic identified by Kobasa (1979) as

> ...the personality characteristic that enabled individuals to remain healthy even when confronted with stressful life events or a stressful environment. The hardy person was someone who recognized that life required him or her to use judgement and make good decisions (control), to become actively involved with others in various activities of life (commitment), and to perceive changes as ultimately beneficial to personal development (challenge) (Pollock, 1989, p. 45).

Hardiness was evident in the way they lived their lives and their attitude to illness. Having had the disease all their lives means they are used to solving problems created by the illness and continuing an active involvement in life. In the words of one participant:

> *Every step along your life things have changed, the disease has always changed...you've always just had to deal with it all the time so you're quite used to doing that.*

The theme of *getting on with life* confirms the finding of others (McWilliam et al., 1996; Lindsey, 1996) that the chronically ill may have a very different way of looking at health and illness from the traditional disease-focused approach. If nurses are to be really effective at supporting the chronically ill then a greater understanding and adoption of this view is required, with subsequent changes in their attitude to appropriate sick role behaviours and compliance with treatment regimes. "Should the goal of treatment be for compliance, as assumed by health professionals, or should it be for a satisfying and optimal adjustment to living with chronic illness as defined by the patient?" (Roberson, 1992, p. 24) Health professionals need to understand the difference in perception they may have from the chronically ill who may see themselves as healthy or relatively healthy. They need to take time to understand the person, their lifestory and the psychosocial aspects of the illness.

Fighting to get on with life

Fighting to get on with life describes the strategies the participants used to cope with the increasing demands of their illness and to maintain their lives as purposeful. The subthemes are:

Redefining what is important

Envisaging a future

Fighting to maintain a positive attitude

Strategies the participants used to live with their illness such as *redefining what is important*, *envisaging a future* and *fighting to maintain a positive attitude* have similarities to coping strategies identified by Craig and Edwards (1983) when adapting to any chronic illness. e.g. seeking relevant information and using intellectual resources effectively, utilisation of hope, preparing for an uncertain future and preservation of a sense of control.

Suffering has been defined as a loss of self that results from leading restrictive lives, experiencing social isolation, being discredited and burdening others (Charmaz, 1983). None of the participants were suffering as a result of their CF. But some participants indicated that they had at times felt threatened by a loss of self because of the illness. They were all leading meaningful lives. Living with a life limiting disease leads them to have wisdom about what is important in life. Younger (1995) describes the development of wisdom as knowing

> "I may lose at any moment, through the play of circumstances over which I have no control, anything whatsoever that I possess, including those things which are so intimately mine that I consider them as being myself" (Weil, 1969). To be aware of this fact in the depth of one's soul is to experience non-being...It is also an acceptance of self that is a willingness to live fully one's own life, to make that life meaningful through acceptance of, rather than detachment from, all that it may hold of both joy and sorrow (p. 65).

Gullickson (1993) describes realisation of our non-being as leading us closer to "the authentic self [which] is aware of the significance of being, which is that we are all being-toward-death...to live is to be dying. Chronic illness brings us into nearness with this understanding" (p. 1388-1391). To live with the knowledge of our own death means that we live well and with an increased understanding of what has meaning and importance to us. One participant describes not putting things off because of this knowledge:

> *I think a lot of people with CF, I've heard it said, that they just live for the moment or they just want to enjoy the next year or whatever it is. I don't think my horizons are that short, well they're quite a bit longer than that. But I certainly am aware that...I don't like to postpone.*

When the disease becomes more unstable or progressive, nurses can best support the person with CF by helping them to maintain a sense of self, by continuing to acknowledge their individual concerns and by assisting with and modifying treatments so that they can maintain the relationships, activities and interests that make up their lives.

For those who live with CF this study describes some responses to the experience which may be useful as confirming their own experience, explaining the behaviour of others and suggesting strategies or responses that may be useful in the future. As life expectancy improves, more people will live longer with this disease. The expectation that adults will assume all the opportunities and responsibilities of a full adult life will increase.

This study adds to our understanding of chronic illness by describing the experience of adults who have had a specific chronic illness all their lives. This is a different experience from that of people who are diagnosed later in life and suggests that, in some ways, it may be easier to know you have a

disease all your life than to be diagnosed later in life "when the structures of everyday life and the forms of knowledge which underpin them are disrupted" (Bury, 1982, p. 169). The fact that this study found like other studies (Shepherd et al, 1990; Walters, Britton & Hodson, 1993) that adults with CF are leading full and active lives that are purposeful and meaningful raises the question of whether prenatal screening, particularly of general populations, is warranted (Friend, 1990).

LIMITATIONS OF THE STUDY

CF is a specific disease affecting a small numbers of adults in New Zealand. This is a study of the personal response of four of those people. It is the voice of the lived experience rather than the hypothesised generalised experience. A limitation of the study is that the need to maintain anonymity meant that full individual narratives could not be described. They would have added richness to the description of the lived experience. Instead this study gives a composite picture of the individual patterns of experiences of the four men and women who participated.

This study has been confined to the experience of adults with CF. It does not include any perspectives (outside the existing literature) of their families, partners, nurses or other health professionals.

SUGGESTIONS FOR FURTHER RESEARCH

Many aspects of the experience were outside the scope of this study including the experience of infertility, bringing up children, relationship to spouse, socio-economic impact of illness and the progressive phase of the illness. These would benefit from further research.

During the study many issues arose which suggest further topics for research. These include: the impact of hospitalisation for an acute episode

on people with a chronic illness, the impact of case management on the experience of people with a long term progressive illness, the different experience of diagnosis at an early age from being diagnosed with a chronic illness later in life, and how adolescents and young adults learn to live with a chronic illness. Finally, each of the themes and subthemes remain tentative and are worthy of further development.

THE RESEARCH EXPERIENCE

Undertaking this research has given me a better understanding of CF, the experience of living with CF and chronic illness. It has confirmed my belief that nurses need to understand patients' perspectives and incorporate them into their practice. The phenomenological approach used in this study was extremely useful when the data took me in a different direction from the one I thought it would. Rather than focusing on the social aspects of an illness, I found myself thinking a lot about mind and body in illness and how this relationship was always present although it was different at different phases of illness. Most importantly, this study and these participants have taught me as a nurse that "health...of itself has no large or notable value...[it] is the fullness of life...with which we have to be most concerned" (Lieb, 1976, p. 169).

It has been a privilege to meet with these four people and to share some of their experience. I am grateful that they were so willing to share with me. I hope that others will find my interpretations of their stories of value.

CONCLUSION

From this study, in conjunction with the existing literature, we learn that these adults with CF gradually conceive their disease as real and that they learn to live with the disease largely through their own experiences. They emphasise getting on with life rather than their disease and this leads them

to minimise the disease to themselves and others, to modify treatments to fit into their lives and to reject the patient role expected of them in hospital. As the disease becomes more progressive they develop strategies to prevent their lives and themselves from being overwhelmed by the illness.

Families and health professionals need to accept that responses to the disease that appear to be denial or minimisation may be appropriate. People with CF are better served by health professionals who have an understanding of them and who can provide continuity of care. The provision of more treatments at home may avoid some of the disruption and hindrance through inactivity presently caused by hospitalisation. Nursing, when considering the increasing numbers of chronically ill, needs to be creating systems of care delivery which enable more continuity. The goal of treatment for people with CF should be "an optimal adjustment to living with chronic illness as defined by the patient" (Roberson, 1992, p. 24).

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APPENDICES
INFORMATION SHEET

THE LIVED EXPERIENCE OF SOME ADULTS WITH CYSTIC FIBROSIS

Who is the researcher?
Pam Doole, Registered Comprehensive Nurse, B.A., Post Graduate Student, Department of Nursing and Midwifery.
Where can she be contacted?
(Telephone number and address provided)
Who is supervising this study?
Judith Christensen PhD, Department of Nursing and Midwifery, Massey University (Telephone number provided)
What is the study about?
The study is trying to describe and understand your experience as an adult (or family member) living with cystic fibrosis and then to make recommendations about the way that nurses and other health professionals could respond to your experience to meet your needs.
What will the participants have to do?
Take part in an unstructured interview describing your experience. It may be necessary to interview you more than once to clarify the meaning of what you have said or to check different experiences that are revealed during the study. The interview will be tape recorded and you can stop the recording at any time. The interview will be transcribed and the transcript taken back to you so you can agree to it being used as data.
How much time will be involved?
The first interview will be approximately two hours. Subsequent interviews may be shorter.
Family members
It may be relevant to interview a family member about their perception of your experience. Your consent would be sought before a family member is approached.
What can the participants expect from the researcher?
To share in the research process and the discovery of the meaning behind your experience. To be kept informed of the progress of the study and to see the completed results.
If you take part in the study, you have the right to:

- refuse to answer any particular question and to withdraw from the study at any time
- ask any further questions about the study that occur to you during your participation
- Provide information on the understanding that it is confidential to the researcher. All information is collected anonymously but because of the small number of adults with cystic fibrosis it may be possible for people who know you to identify you in any reports that are prepared from the study, although all identifying details will be changed. The information produced will be used for the thesis, in published reports, written and oral presentations of the research, ongoing reports to students in the thesis group and in discussions with my supervisor.
- Be given access to a summary of the findings from the study when it is concluded.

If you have any concerns about the research you may write to the Chairperson CRHA Wellington Ethics Committee, Wellington Hospital, Private Bag 7902, Wellington South.

CONSENT FORM

THE LIVED EXPERIENCE OF SOME ADULTS WITH CYSTIC FIBROSIS

I have read the information sheet for this study and have had the details of the study explained to me. My questions about the study have been answered to my satisfaction, and I understand that I may ask further questions at any time.

I also understand that I am free to withdraw from the study at any time, or to decline to answer any particular questions in the study. I agree to provide information to the researcher on the understanding that every effort will be made for confidentiality to be maintained as stated in the information sheet and my approval is sought before the transcript is used as data.

I understand that the researcher will seek consent from me before approaching any of my family members to be part of this study.

I wish to participate in this study under the conditions set out on the information sheet.

I understand that the procedures have been approved by the Central Regional Health Authority Ethics Committee.

Signed: _____

Name: _____

Date: _____

I consent to the researcher, Pam Doole,

approaching _____ (family member) to be involved in this study.

Signed: _____

Name: _____

Date: _____

Judith Christensen, Department of Nursing and Midwifery, Massey University (telephone number) is supervising this study.