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Fighting for the breath of life: A sociological study of patients with cystic fibrosis

Narcez, Louis Howard, Ph.D.

City University of New York, 1989

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FIGHTING FOR THE BREATH OF LIFE: A SOCIOLOGICAL STUDY OF PATIENTS WITH CYSTIC FIBROSIS

by

LOUIS H. NARCEZ

A dissertation submitted to the Graduate Faculty in Sociology in partial fulfillment of the requirements for the degree of Doctor of Philosophy, The City University of New York

1989

This manuscript has been read and accepted for the Graduate Faculty in Sociology in satisfaction of the dissertation requirement for the degree of Doctor of Philosophy.

Feb 10, 1989
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M E Brown for Patricia Kendall
Chairman of Examining Committee

Feb 10, 1989
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Abstract

FIGHTING FOR THE BREATH OF LIFE: A SOCIOLOGICAL STUDY OF PATIENTS WITH CYSTIC FIBROSIS

by
Louis Narcez

Adviser: Professor Patricia Kendall

This dissertation examines the plight of the patient with cystic fibrosis. It is based on short interviews with cystic fibrosis patients and health care providers, and on observations in a hospital setting. The dissertation draws upon the sociology of medicine, the sociology of death and dying, and on the sociology of deviance.

ACKNOWLEDGEMENTS

This dissertation would not have been possible if it were not for the undying dedication of the members of my Ph.D Committee: Dr. Patricia Kendall {Chair}, Dr. Charles Winick, Dr. Michael E. Brown, and Dr. Lindsey Churchill.

All of the members of my Ph.D Committee have known me for a long period of time. I was a student in their undergraduate and/or graduate courses, conducted research under their aegis or I was their research assistant. They are owed a debt of gratitude for being generous and sharing with me their precious time and vast knowledge and expertise.

Recognition is also due Dr. Melvin Reichler, of Queens College, who introduced me to the world of computers, and who offered many helpful suggestions pertaining to the appearance of this dissertation. His course in Sociological Analysis, taken over a decade ago, has influenced my research activities ever since.

I am grateful to Dr. Renee C. Fox. Her seminar in the Sociology of Medicine, taken while I was a graduate student at the University of Pennsylvania, was the catalyst that ignited my interest in the sociology of medicine and health care.

Special thanks are due my younger sister, Phyllis Narcez, who offered many constructive and valuable suggestions on draft versions of the chapters.

I also wish to acknowledge the late scholar and humanitarian, Dr. Edward Sagarin. His seminar on Deviant Behavior thoroughly addressed many concepts and ideas which proved very useful in connection with my research.

Writing this dissertation proved to be a very painful experience --- far more painful than most people could possibly imagine. In the course of this venture, four persons who were essential to its success, passed away from cystic fibrosis. There were times when I felt certain I was doing a dissertation in the sociology of death and dying, rather than in the sociology of medicine.

I am grateful to Bill { pseudonym }, the youth life specialist on the adolescent unit, who first suggested that I might consider doing a dissertation concerning cystic fibrosis. It was through him that I was introduced to a number of patients with cystic fibrosis. We spent many nights together in the hospital lounge, consuming Campbell's clam chowder, and discussing ideas related to this dissertation.

Undoubtedly, my greatest debt is owed to the CF patients who shared with me their feelings, ideas, and perspectives. Without their assistance, this dissertation would never have materialized. It is my sincere hope that this dissertation does a sense of justice to them, and to all victims of the life threatening, vicious disease, cystic fibrosis.

Louis H. Narcez

In Memory and Honor of my father
Arnold Narcez { 1929 -1983 }
who is in the Heavens

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Chapter 1: BACKGROUND TO THE PROBLEM

Introduction:

Cystic Fibrosis [CF] is an inherited and lethal disease that affects one in every one thousand live births in the United States. The disease is also common elsewhere in the world including England, Canada, Australia, New Zealand, and in Europe. Most victims of the disease are Caucasians. The disease also affects Blacks, but seldom Orientals.

The disease is characterized by the inability of the exocrine glands [glands that produce mucus, saliva, and sweat] to function properly. Although the cause of the basic defect is unknown at the present time. it is thought by some scientists that a " key enzyme " responsible for the proper transport of sodium and chloride is defective or inoperative.

The disease affects the respiratory and digestive systems of the body. Due to the absence of significant [normal] levels of pancreatic enzymes, the patient is unable to thrive, gain weight, or acquire an adequate level of nutrition necessary for survival. It is, however, the respiratory complications which

are most often life threatening. As the disease progresses, pneumonia, atelactasis, broncheictasis, and pneumothoracies occur, and often recur. Frequently, due to the pulmonary complications, heart damage [cor pulmonale] is also present. Diabetes, liver, kidney and other types of damage frequently develop in cystic fibrosis patients. Death from cystic fibrosis usually results from the pulmonary damage and complications associated with the disease.

Before the mid 1950's, few CF patients lived past the early years of life. Treatment for the disease was clearly in the " dark ages ". Many of the treatments in use today such as antibiotic therapy, pancreatic enzyme replacement, vitamin and nutritional supplementation, and chest physical thearpy were not discovered at this point in time.

In 1956, intensive prophylactic therapy was begun in Cleveland, Ohio. The treatment plan was geared towards preventing further complications affecting the lungs as well as the digestive system. This preventive therapy consisted of the following elements:

{1} physical therapy: postural drainage of the

patients lungs.

{2} inhalation therapy: patients were given medications to loosen mucolytic secretions.

{3} antibiotic therapy : to combat respiratory infections, patients were given antibiotics orally, intravenously, or nebulized into the patients lungs.

{4} Pancreatic extract: to aid in the digestion of food, pancreatic enzymes were prescribed.

{5} diet: patients were put on diets which were high in fat soluble vitamins, high in caloric intake, and low in fat. [Waddell: 1985]

Since the 1950's, the life span of patients with cystic fibrosis has increeased somewhat. The disease is still the leading cause of congenital death in children. Although the life span of patients with cystic fibrosis is about twenty (20) years of age [National Cystic Fibrosis research Foundation], greater numbers of patients are surviving through the early teen, young adult, and adult years. Nevertheless, treatment of the disease is still regarded as palliative [Schwachman: 1980], and many patients with cystic fibrosis die during the early weeks or months of life.

Popularity:

Unlike other diseases, cystic fibrosis does not

experience the recognition of cancer, epilepsy, or diabetes. One reason is that there is a lack of a key figure in the general publics view who is associated with CF. For example, President Kennedy has a sister who is mentally ill; Harvey Firestone has a son who is mentally retarded. To date, no celebrity has a child who has cystic fibrosis. Jerry Lewis has a long standing tradition of raising funds for muscular dystrophy, and the general public often identifies victims of muscular dystrophy as " Jerry's Kids ". Although, in the past few years, Hugh Downs and Joan Rivers have served as chairpersons for the National Cystic Fibrosis Research Foundation, there efforts were relatively short lived.

Another indication of the popularity of a disease is the extent to which it is reflected upon in written articles. According to Index Medicus, the number of articles written about cystic fibrosis in medical journals for the past ten years is as follows: 1988-393; 1987 - 458; 1986 - 474; 1985 -415; 1984 - 369; 1983 -360; 1982 -329; 1981 - 298; 1980 - 322; 1979-320; and, 1978 -265. It is interesting to note that since 1986, the numbers of articles on CF appears to

be declining. In addition, the vast majority of articles on cystic fibrosis deal with biochemical and medical aspects of the disease rather than addressing psychological or behavioral aspects of the illness. For example, In 1987, there were 458 articles but only 4 concerned nonmedical aspects of the disease.

In nonmedical publications, cystic fibrosis is seldom given sweeping attention. According to the Readers Guide to Periodical Literature, the number of articles appearing in popular magazines concerning CF is as follows: 1988 - 4; 1987 - 6; 1986 - 7; 1985-3; 1984 - 3; 1983 - 3; 1982 - 1; 1981 - 3; 1980 -4; 1979 - 4; and 1978 - 4.

Studying CF Sociologically:

Medical sociologists hve a long standing interest in issues related to medicine and health care such as: the quality and economics of health care, the social dimension of illness including the effects of illness on social and familial relations, new trends and advances in the treatment and management of disease; and the allocation and dispersement of medical resources and technology. In addition, a growing number of med-

ical sociologists have a strong interest in the areas of catastrophic illness and in the "sociology of death and dying." As the cost of medical care continues to rise accompanied by radical and momentous advances in technology, it appears inevitable that these concerns and issues will continue to dominant as key concerns of medical personnel and medical sociologists.

Cystic fibrosis is the most frequent severe genetic disease of childhood in the United States, for which there is no successful treatment 1 Although, there is no cure for cystic fibrosis, there is a regimen of palliative care which most patients follow in an attempt to improve their life span.

When cystic fibrosis was first described and discovered by Dr. Dorothy Anderson, there was virtually little understanding of the disease and there was absolutely nothing that remotely resembled

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1 Zigler,E. and Stevenson, M, CHILDREN: Development and Social Issues, Boston, Mass., Health Publications, 1987, p. 211

"treatment" or management " of the disease. 2 Gradually, as the knowledge of medicine and science has expanded, so has our understanding of cystic fibrosis. With the development of new antibiotics and microbial agents, new bronchodilators, and new techniques in chest physiotherapy, the prescibed medical protocol for cystic fibrosis has greatly improved. This expansion in the medical management of cystic fibrosis has resulted in sharp increases in the cost of providing care to cystic fibrosis patients.

Although there is no cure for the disease, changes in the character of treatment have yielded an increase in the longevity of some CF patients. As is the case with other catastrophic illnesses, the medical costs involved are often exorbitant and adversely affect the financial status of the patient.

Cystic fibrosis patients are also of interest to sociologists because they provide primary data for studying the social character of illness, and the impact that life threatening illness has on one's

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2 I am indebted to a physician {pediatrics} who provided me with this information.

position in society. Sociologists and social psychologists such as Goffman, Parsons, Freidson,and Lemert, have discussed in detail the " stigmatization processes " and reaction which illness solicits from other members of society. Patients being treated for catastrophic illnesses, such as cystic fibrosis, can be analyzed and studied as "involuntary deviants" in a society where conformity and a lack of tolerance towards deviants { be they voluntary or involuntary } are the norms.

This dissertation will focus on the plight of the cystic fibrosis patient with emphasis on the social and psychological character of the disease, the everyday realities of living with a life threatening disease, and how cystic fibrosis patients strive to maintain a sense of "normality" in spite of the fact that from a sociological perspective they are considered "deviants."

Chapter 2: METHODOLOGY

The data in this dissertation was gathered by means of employing a variety of research methods including participant observation, the " snowball method " of social research, and interviews with CF patients and health care providers.

The Patients:

Interviews were conducted with 50 cystic fibrosis patients and members of their families { parents, girl- friends, siblings, etc. }. The patient sample consisted of approximately 70% males { N=35} and 30% females { N = 15 }. The vast majority of patients interviewed were over 21 years of age. My decision to rely more heavily upon persons over age 21 was based on the following considerations: {1} I felt there was a greater likelihood that older patients would prove more credible subjects. It was my impression that young adults were likely to reflect upon their situation with less exaggeration than younger persons. {2} A person is of legal age when they reach 21 years old. Therefore, there is no need to secure the consent and permission to quote anything that the

subject might offer. On the other hand, if I had opted to interview minors, whether " informed consent " should be obtained would be an ethical issue warranting deep consideration. Moreover, I would most likely feel very uncomfortable asking a parent for " informed consent " to interview their son or daughter. There is always the danger that questions I might ask might prove upsetting or beyond the child's grasp and understanding.

In cystic fibrosis, the standard by which the severity of the disease is measured is the **Schwachman - Kulczycki Scale.** The scale takes into account the following: {1} pulmonary status of the patient { ie, whether the lung fields are clear or whether rales and/or rhonci are present; the degree to which hypoxia and dyspnea are present; the degree of permanent lung damage that is present { interstitial fibrosis, other lung changes found that are associated with emphysema, etc. }, and scarring caused by pulmonary infections and pnemothoracies ; {2} the gastrointestinal involvement {ie, the deficiency of pancreatic enzymes, and other GI symptomology,} and {3} other factors including the patient's ability to cope and thrive, and the

extent of his/her limitations { eating, walking, GI pain, etc.}. The patients whom I interviewed varied greatly in terms of the progression and severity of their disease. Some patients in the sample represented mild cases of CF in terms of the Schwachman-Kulczycki Scale. Other patients were more advanced cases of cystic fibrosis, and a few patients were at the final stage shortly approaching death. All of the patients I interviewed were " regulars " to the hospital scene. They were familiar to the members of the nursing staff at the hospital. { This is discussed in detail in Chapter 5 }.

Since 1978, I have served on an intermittent basis as a hospital volunteer. This allowed me an opportunity to come in contact with a number of CF patients. As time progressed, my ties and familarity with various patients became stronger. Since CF patients are usually hospitalized on a regular basis, I came in frequent contact with many of the patients. In terms of my research for this dissertation, this proved beneficial. Since the patients as well as myself knew each other for a long period of time, neither party felt uncomfortable in their interactions

with the other.

In 1983, the hospital hired a young man of thirty years of age to serve as a youth - life specialist . This person, Bill {pseudonym} was responsible for engaging hospitalized patients into recreational activities. I established a very friendly and professional relationship with this person. Bill introduced me to a few CF patients.

Medical Personnel:

I also engaged in relatively short discussions and interviews with medical care providers who rendered care to CF patients. Included in my pool of medical personnel were nurses, interns and residents, three physical therapists, and a pharmacist.

Registered Nurses:

Most of the health care providers I interviewed were registered nurses. Generally, I found that nurses were likely to talk openly about their role in caring for CF patients and about their personal feelings. The nurses that seemed more talkative and approachable tended to be people who knew

me for some length of time, or younger nurses who recently graduated from nursing school.

I found that the best time to gather data from nurses was during the following: {1} late evening when visiting hours were over and when patients needs for care were at their lull point; {2} on weekends, especially early mornings such as Saturday or Sunday around 10 AM - 11 AM. The floor tended to be quiet or deserted at this time. For example, the interns and residents made " quick rounds " on Saturday mornings, and on Sunday mornings the house staff did not conduct formal rounds. Most private physicians seldom visited their patients on Saturday, and virtually none ever showed on Sunday mornings. { The only exceptions appeared to be the CF team and the pediatric oncologists caring for cancer patients. The doctors of the CF team always made rounds on both Saturday and Sunday mornings }. When fewer persons were around nurses seemed to talk relatively freely. {3} While I was visiting CF patients and nurses were in the patients room. An ideal situation for gathering data was when nurses were performing postural drainage on the patient. Since the drainage session usually lasted a few

minutes, nurses frequently spoke during the procedure.

Physical Therapists:

I found that physical therapists often talked about the nature of their work. In fact, two physical therapists that I spoke with not only talked about postural drainage but they also talked about other topics as well. One physical therapist frequently engaged in conversations as to why she should be allowed to treat patients without a doctor's referral. The other physical therapist often spoke of the competition and ongoing debate between their profession and respiratory therapists as to which group of professionals could best provide postural drainage to CF patients. In her view, respiratory therapists should continue to administer oxygen and aerosol medications to CF patients while leaving the " heavy work " { postural drainage } to physical therapists. According to her, physical therapists have a greater understanding of physical anatomy and they have greater experience with patient contact than respiratory therapists.

A third physical therapist I spoke with talked extensively about cystic fibrosis patients as well as

the vital need for CF patients to " religiously " perform drainage at least twice daily. She remarked that some younger CF patients perform postural drainage when they are not feeling well but, once they start feeling better they tend to neglect this facet of treatment. Her feelings on this subject were echoed by two or three nurses I interviewed.

Techniques:

In organizing the material to be addressed in this dissertation, I decided to draw heavily upon the method that Goffman used in his work Stigma. I identified a series of areas { which represent each of the chapters in this dissertation } which I systemically tried to address with respect to the patients. For example, I chose to explore concepts and ideas in sociology such as Parson's " sick role" model, staff-patient relations, the everyday realities of life with cystic fibrosis, etc. I memorized series of questions related to each of these areas and I routinely asked patients to offer there input on such matters.

The first few subjects whom I interviewed agreed to be tape recorded. After only two interviews, I

chose for a variety of reasons to abandon this practice. While the majority of patients indicated a willingness to be taped, a number of nurses and other medical providers did not seem happy about the idea. I came to the conclusion that there were many drawbacks to this approach. For one thing, in the back of my mind was the feeling that subjects may be hesitant to say things freely if they know that they are being taped. I also felt that some subjects might say things that they may later decide seemed inappropriate or prefer not to have stated. { I did not want to place people in a position of worrying about anything they may have said }. Most of the patients I encountered did not seem to mind that I would write things down { I took notes during my interactions in a small 5 X 8 stiff back memo pad } in their presence. In fact, one patient remarked something to the effect that he was glad that I was taking notes so that I could accurately record what he was saying. In light of these considerations, use of a tape recorder was not feasible.

Role Models:

In the course of my fieldwork, I discovered that

there were two CF patients who were greatly admired by other CF patients. Both of the individuals were viewed as " role models " because they were in their early thirties, highly educated, and, in spite of their illness, they held prestigious jobs. { One person was a business executive, and the other was a senior physicist. }

Hospitalized CF patients often congregated around Jack and Lloyd { pseudonyms }. I often engaged in conversation with them. I found that my relationship with both persons, was of enormous benefit in terms of my being able to relate to other patients. A number of patients viewed me as a friend of theirs and consequently they spoke freely and candidly in my presence.

Importance of People:

When conducting research with human subjects, it is essential to concede the benefit that the participants provide. Bearing this in mind, I was extraordinarily careful to make my research subjects feel important as persons as well as subjects in my study. I tried to instill in my subjects the feeling that I

was just a fellow human being, and in no way strikingly different from them. I was very careful not to be judgmental or to formulate opinions concerning remarks that were stated.

At the end of each interview, I asked every person if he/she would like me to emphasize anything in particular when I write my thesis. Few people offered any pressing suggestions. However, several hospitalpized patients asked me to praise the dietitian, Tenaj { pseudonym }, for her extreme dedication and caring. { This is discussed in Chapter 5 under the subtitle, " Beautiful People " }.

Chapter 3: ILLNESS AS A DEVIANT STATUS

Parsons's "Sick Role"

Talcott Parsons [1951] has suggested that sickness in and of itself and under all conditions is a deviant status. 1 Parsons' position is based on the assumption that there is a debilitating component of illness which interferes with normal everyday functioning. Some scholars have serious questions about this assumption. The limitations experienced by an individual because of his/her illness is highly variable. Certain illnesses or disabilities undoubtedly are more debilitating than others. For example, as Parsons notes, " The diabetic takes his insulin or other medication and pays some attention to diet, but is in every sense socially competent." 2 Moreover, the individual's own personal adjustment to his medical condition varies greatly among individuals. Factors such as patient compliance, readiness to assume responsibility for one's well being, and familial support are some variables of relevance in

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1 Parsons, Talcott, The Social System,{ Glencoe Il, The Free Press, 1951 p. 294 ff.}

2 Ibid, p. 295

this regard. Hence, it is not surprising that some persons stricken with life threatening illnesses are better able to adjust to their status than others suffering from less serious ailments.

Victims of cystic fibrosis are engaged in a constant fight to sustain their lives. This does in fact require strict adherence to tedious medical protocols. Nevertheless, I encountered some cystic fibrosis persons who, in spite of their illness, were able to hold down jobs and satisfactorily function like most other members of society. Among the CF people I interacted with were individuals who worked in jobs requiring higher education, talents and/or skills. A number occupied business positions; some served in less strenuous occupations which largely entailed work of a sedentary nature (bus dispatcher, business executive, telephone clerk, secretary, bookkeeper, accountant in a large firm, computer analyst, etc.) One CF patient held a lucrative position as a highly paid consultant for a government agency. A number of the older patients held supervisory or managerial positions. One male subject served as a unit supervisor for a federal government agency. One particularly intelligent per-

son (recently deceased) was a senior research scientist for a private firm on Long Island. This individual (a physicist) was highly regarded by his colleagues. He had published numerous articles in prestigious scientific journals. He was so talented and ingenious that he was able to build his own computer, before computers were widely used. In college, he quickly " rose to the top " and was elected to the scientific honorary society at Queens College 3 This person does not represent an isolated case. In the course of conducting research for this dissertation, I was frequently introduced to CF patients who triumphed academically, or who accomplished admirable feats, in spite of their physical limitations.

At a recent meeting outlining major advances in research on cystic fibrosis, the key speaker, an executive director of medical affairs for the National Cystic Fibrosis Research Foundation, noted in passing, that some of the leading scientists in CF research include a biophysicist in North Carolina who has cystic fibrosis, and a biomedical researcher in Calif-

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3 Obituary in **NEWSDAY**: The Long Island Newspaper 28 November 1986, p.38

ornia who also has cystic fibrosis. Interestingly, when the speaker made this remark, a few cystic fibrosis patients said, " It would probably take a fellow CF patient to find a cure ."

One of the expectations associated with Parson's sick role model is that " the individual is excused from social responsibility." 4 While I would prefer to refrain from making categorical generalizations about any group of research subjects, it was my impression that most of the CF patients I met seemed to be self conscious, accomplishment oriented, and virtually all of the older persons seemed to have a sense of purpose or direction. For example, one male patient spoke of his desire to become a college professor, while a female patient talked exyensively of her desire to excel in the field of fashion design. There are already cystic fibrosis patients who have become physicians in New York, California, Philadelphia, and Texas. The CF patients I encountered did not seem to look for an excuse to avoid social responsibilities as far as working or other activities

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4 Parsons, Talcott, op cit., p. 294 ff.

were concerned.

The fact that a number of CF patients can attend to their medical needs (a vigorous treatment plan in virtually all cases) and simultaneously carry out meaningful work and/or productive activities, stands as a testament by itself. Schwachman's study [1974] of 70 CF patients over 25 years of age found that 28 were married, 40 held bachelor's degrees, 13 held master's degrees, one was a medical doctor, two were lawyers, three were nurses, one was a physical therapist, and one was an engineer. 5 It would appear that Parsons' component of the " sick role" concerning relief from social responsibilities is more applicable earlier in the case of acute illnesses, as opposed to the situation of chronic illness of all CF patients.

Nevertheless, there are a few cases in which Parsons' insight bears relevance to cystic fibrosis. However, this usually concerned CF patients who were quite ill, at the later stages of the disease, or in-

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5 Schwachman, H., Kowalski, and K.T. Khaw, " 70 Patients with Cystic Fibrosis over 25 years of age--a new Outlook." **Cystic Fibrosis Club Abstract**, 1974, 13:3

capacitated by cystic fibrosis in addition to simultaneously suffering from other diseases. A young woman (now deceased) provided the following insights:

> " Poor health has deprived me of both the big and little pleasures that most people take for granted. Such routine tasks as shopping and getting dressed are incredibly difficult or impossible. I walk slowly and can't stand for long periods of time. Even hugging someone is now painful to me.....6

This young adult experienced grave hardship and incapicities because of advanced lung disease from cystic fibrosis, coupled with crippling rheumatoid arthritis.

Parsons' formulation of the " sick role " also includes three other expectations. These are: (1) the individual is seen as not at fault for his/her illness. (2) The condition is defined as undesirable, and, (3) The individual has an obligation to seek technically competent medical help. 7 These expectations associated with the Parsonian model are applicable to cystic fibrosis. Since there is nothing that an individual can do to bring about this illness,

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6 Donovan, Elizabeth, " Viewpoint " in Glamour Magazine, July 1984, p. 30.

7 Parsons, ibid

the person with cystic fibrosis is not at fault for having the disease. Cystic fibrosis is viewed as an undesirable condition especially in view of the fact that CF is a serious life threatening disease which has few, if any, " secondary gains. " 8 Individuals with CF, although usually responsible for taking care of their everyday medical needs, require the assistance and services of physicians and ancillary medical personnel.

Szasz-Hollender Model:

Szasz and Hollender [1956], both of whom are physicians, examined the relationship between behavioral implications and illness. They identify three models of the doctor - patient relationship.

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8 One of the few "secondary gains" might be the absence from school when extremely inclement weather occurs. For adults there are few secondary gains. In spite of the seriousness of the illness, CF people do not benefit from compensation for their illness. The governments attitude towards CF patients is one of indifference. Due to DRG classification, CF is regarded as a gland condition, and not as a pulmonary disease of life threatening classification. This saves the government a great deal of money at the expense of CF patients.

These include: (1) activity - passivity, (2) guidance- cooperation, and, (3) mutual participation. 9 In the " activity - passivity" model, the physician is primarily responsible for caring for the patient. The patient's role is limited to being the recipient of a practitioner's care. The second model which Szasz and Hollender identify is the " guidance-cooperation model. In this model, the physician instructs the patient as to what needs to be accomplished. Therefore, the physician instructs the patient as to what he/she must do, and the patient is responsible for carrying out the physicians orders. This model is applicable to situations in which a person might be ill with an infection and the doctor prescribes bedrest and medication; the patient is expected to comply. The third model which Szasz and Hollender present is the " mutual- participation" model. In this model, the role of the physician is to help the patient to help him/herself. In this model there exists a partnership between the physician and

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9 Szasz, Thomas & Hollender, Marc, " A Contribution to the philosophy of medicine: the basic models of the doctor-patient relationship, " A.M.A. Archives of Internal Medicine, {May 1956}, vol 97: 585-592.

his/her patient. The " mutual participation" model is most appropriate to clinical situations involving chronic illness such as cystic fibrosis.

In the course of conducting interviews with CF patients, virtually all indicated that they maintained very close contact with their physicians. For example, one of the physicians in the CF Center, insisted that all of his patients maintain daily phone contact for at least a week, following discharge from the hospital. Patients were expected to call this doctor every morning before 8:30 A.M. at his private office. 10

One indication of the " partnership " that exists between CF patients and their physicians is the communication that exists between the patients and the health care providers. Patients are instructed as to situations that might conceivably arise, and how to deal with them. Patients are informed that certain situations are of major importance. For example, if a

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10 The only day they were not expected to call was on Sunday. Some patients stated that this doctor would call them before they had a chance to reach him.

patient has hemoptysis (coughing up blood, or blood in sputum), he/she is advised to call the doctor and to plan on being seen in the emergency room. Patients are told that when they are bringing up blood, they should note the amount (teaspoon, tablespoon, etc), and the character of the blood (ie, is it dark red or bright red, is it blood alone that they are coughing up, or is the blood in streaks). Information of this nature is very important since it provides the physician with information as to the source of the blood, and its possible cause. Similarly, patients are informed that sudden chest pain, a feeling of pressure or of a leaning in the chest, or excessive coughing and/or dyspnea (difficulty breathing or shortness of breath) , or a fresh infection accompanied by fever, loss of weight, are also situations which warrant immediate medical attention.

Another indication of the mutual cooperation that exists between the doctor and the patient concerns the credibility physicians attached to the patients own accounts of their status. Physicians would routinely asks patients whether they felt better with some medications as opposed to others. Questions such as " Do

you feel better receiving Keflex and Tetracycline orally or Augmentin and Tetracycline ? " or " How does Dicloxacillin get along with you-- does it cause you to have loose stools ?", were routinely asked of patients. In the hospital setting, physicians would sometimes ask a patient " How are you tolerating the Promaxim ? Is it giving you nausea ? Can you handle it ?" If patients indicated that they felt very uncomfortable while maintained on one medication, or if they noted that they felt better with one combination of antibiotics as opposed to another, the CF physicians would generally give this some weight in considering whether to change the patients' dosage and/or medications. Based upon all of these factors, it would seem that Szasz and Hollender's model of " mutual participation " applies to cystic fibrosis patients.

Freidson's Typology:

Eliot Freidson [1970] discusses the societal reaction to various types of deviance, including illness. Freidson maintains that societal reactions depend on whether the person is held responsible for

the condition and its seriousness. 11 Freidson views illness in terms of whether it represents a " minor deviation " or a " serious deviation ." Friedson offers the following typology:

Table 2-1: Types of Deviance, by Quality and Quantity of the Societal Reaction

Judgment of seriousness	Judgment of Responsibility	
	Ind held Resp.	Ind. not held Resp
Minor deviation	slight addition to obligations; minor or no suspension of a few privileges or a fine {for example a parking violation}	Partial suspension of a few ordinary obligations; slight enhancement of privileges; obligation to get well { for example, a cold}
Serious deviation	Replacement of ordinary obligations, by new ones: loss of privileges {for example, murder}	Release from most ordinary obligations; addition to privileges; obligation to seek help and cooperate with treatment {for example, a heart attack}

Based upon Freidson's typology, cystic fibrosis patients would most like fall into the category representing individuals with a serious deviation, in which the individual is not held responsible for his or her condition.

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11 Freidson, E., Profession of Medicine, { New York: Harper & Row Pub., 1970, pp.230 ff. }

Stigmatization:

In his classic work, Stigma , Goffman addresses the social situation of stigmatized individuals. Stigmatized individuals are unable to conform to the standards that society considers normal, persons who are disqualified from full social acceptance, or persons who must strive to adjust their precarious social identities. Included in Goffman's framework of the " stigmatized " individual are some physically ill people. Some of the concepts which Goffman identifies are relevant to the everyday lives of some CF patients.

A person who appears ill is more likely to be stigmatized than a physically ill person who shows no outward signs of his/her illness. As Goffman notes:

> " When an individual's stigma is very visible, his merely contacting others will cause his stigma to be known about. 12 For example, a participant with a speech impediment,... can hardly open his mouth without destroying any unconcern that may have arisen concerning his failing, and he will continue to introduce uneasiness each time thereafter after he speaks

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12 Goffman, Erving, Stigma: Notes on the Management of Spoiled Identity, { Englewood Cliffs, N.J.: Prentice Hall, 1963, pp. 48 ff.}

> .13 Similarly, a blind person with a white cane gives quite visible evidence that he is blind, but this stigma symbol (the cane), once noted, can sometimes be disattended, along with what it signifies. But, the blind person's failure to direct his face to the eyes of his co-participants is an event that repeatedly violates communication etiquette and repeatedly disrupts the feed-back mechanics of spoken interaction. 14

Cystic fibrosis is an interesting disease in that some patients present visible signs of the disease, while others do not. Among outward signs and symptoms of the disease are: (1) clubbing of the fingers, (2) fingernails that are white or appear cyanotic (latter stages-- this signifies a deficiency of oxygen , (3) a " pot belly " (often present in young children and patients with substantial or marked gastrointestinal involvement), (4) a " barrel chest " (a result of pulmonary infections/ respiratory processes), (5) excessive sweat loss (a result of abnormal transport of sodium chloride in intracellular activity), (6) excessive coughing which may or may not be productive in terms of mucolytic secretion production, and (7) shortness of breath.

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13 Ibid, p.48

14 Ibid, p.49

Depending on the severity of the disease, and the degree of visibility of the symptoms, a cystic fibrosis patient may or may not be able to cover or conceal his/her illness from other people. The extent to which the disease is visible is of key importance for a variety of reasons. Many CF people are acutely aware of the disease's signs, and many patients are self conscious and greatly concerned with their self-image. The following sentiments and remarks expressed by some CF patients illustrate this:

> " I also have to contend with looking ill. Sometimes I hardly recognize myself in the mirror and I want to weep with the longing for the way I once was."15

These sentiments expressed by a young woman represent her own acknowledgment of how CF has negatively and appallingly changed her personal appearance. Behind this woman's perception of her self is an intense feeling of sorrow when she reflects back in time to when her physical appearance was untouched by the ravishing signs of her illness.

Another patient says:

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15 Donovan op cit p.30

> " My fingers are badly clubbed. I am very careful to hide my fingers from the immediate visual field of my friends, coworkers, or people I come in contact with."
>
> .....young adult male

This statement reveals an awareness on the part of the individual that his fingers are different from others. To avoid a negative reaction or possible stigmatization, he consciously avoids displaying a visible sign of his illness.

Still another patient points to a different manifestation of CF, and discusses his growing awareness of it:

> " Ever since I was a child, I had a pot belly. When I was young, I did not seem to care about it. Perhaps, it was because other kids I knew also had pot bellies. Now that I am grown, and only I have a pot belly, it has made me very self conscious."
>
> ..... adult male

In his comment, the man indicates that he has always been aware that he has a pot-belly. When he was a child, he wasn't bothered by this fact, because he did not feel different from other children who also had pot bellies. However, as he became older and aware of the fact that now, only he has this vis-

ible sign of CF, he feels differently about the matter.

An adult man suggests that he is always aware of the outward signs of his disease:

> " With a disease like cystic fibrosis, it is difficult to hide your illness from other people. I am always coughing and the slightest activity causes me to be out of breath, and in turn, to cough even more. My chest is long and thin, not very muscular, because of the pneumonias and pneumothorax that I recently had."
>
> ..... adult male

To hide visible signs of their illness, some CF patients have developed effective strategies. For example, one young woman says:

> " Whenever my fingernails turn bluish I immediately put on dark nail polish. Since the nails often become quite discolored, I find that I cannot polish the nails pink, or light red, white, or some light color. I am forced to polish the nails brown, green, dark blue, or some other less pleasing color. Still, it is better to have fingernails that are not particularly attractive (because of the color of the polish), than to have fingernails that are cyanotic."
>
> .....an 18 year old female

Another regulates his social activity according to his ability to act " normally ":

> " I never go out socially when I feel low winded or out of breath. This has resulted in my curtailing my social activities.....But, given the choice of going somewhere, enjoying the day out and being out of breath and feeling ill, or staying home watching the football game on TV, I'd rather stay home".
>
> ..... a CF teenager

The remarks of this teenager suggest that he limits his social activities so as not to reveal symptoms of his underlying disease.

Another patient with cystic fibrosis echoed similar sentiments:

> " When I was in college, I frequently had to miss classes. There were many times when I was really out of breath and felt very fatigued. I almost always arrived late to class. I frequently stopped at the water fountains along the way. There were a couple of days when I was very tight. The profs [sic] in the _____ [name of department] just tolerated me." 16

This person also acknowledges the limitations imposed by being short of breath. In the previous comment the teenager stated that his social life was affected by difficulties breathing. However, in the latter remark, a college graduate notes how pulmonary

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16 The remarks in this statement are paraphrased. These sentiments were told to me a short time ago. The person who made these remarks has since died.

complications of cystic fibrosis caused him to be late to his college classes, and thereby threatened his academic performance.

The awareness of stigmatization arising out of coughing in public was a theme that frequently emerged in conversations with some patients who had cystic fibrosis. An adult CF patient stated:

> " Every person with lung disease must learn to suppress their cough if they intend to keep their job, and hide their illness."

Another stated:

> " To prevent attacks of coughing and shortness of breath you must do three things: 1) master the techniques of suppressing your cough, 2) carry around your Proventil inhaler (a bronchodilator), and 3) drink plenty of coffee... the caffeine in coffee opens up the lungs.... it acts like theophylline (a bronchodilator).
> ..... an adult CF patient giving advice to fellow CF patients at an informal meet

Stigmatization in the Hospital:

Cystic fibrosis patients are not only victims of stigmatization occurring in " the real world" but, they also experience stigmatization within the confines of the hospital setting. This frequently re-

sults because of noticeable traits or characteristics associated with their illness. Patients who are roommates of CF patients sometimes complain that they cannot get any sleep at night because the patient in the other bed was coughing the whole night and it kept them awake. Many CF patients seem to prefer a room which is slightly chilly or cold. Some CF patients claimed that it made them breathe more comfortably. This, too, can create problems in terms of relations with other patients who are hospitalized and sharing the same room. During my tenure as a volunteer, I remember one patient who kept the room freezingly cold. Although the patient was in a private [single] room, what might be termed " mild inconveniences" ensued. Nurses frequently commented that they felt a chill in the room, and they tried to persuade the patient to lower the air conditioning. One nurse who worked late evenings wore three hospital gowns over her dress when she had to enter the room. The room was located next to the patients lounge, and patients who passed by on their way to the lounge would frequently make a remark such as " There's a draft over here."

Treatment of the disease itself often made CF patients become subject to stigmatization and unwelcome attention. For example, some patients require oxygen in order to breathe easily. When oxygen was being used a highly visible sign was placed on the door of the room. The sign read in capital letters NO SMOKING --- OXYGEN IN USE. The sign also had a caution symbol and its presence was unmistakeable. Sometimes visitors in the halls could be hear remarking that " the patient in that room must need that (oxygen) to breathe." Sometimes nurses would leave the door to patients room open while the patient was receiving postural drainage or receiving oxygen via a nasal cannula or venting mask. Some Cf patients reported to me that they found this disturbing or embarassing. Other patients who walked through the hallways while receiving oxygen via an " E tank" also mentioned that they felt awkward, embarrassed, or stigmatized because of their predicament. 17

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17 One physician told me that CF patients, as well as anyone with a pulmonary disease, needs to exercise to maintain cardiac function. It was for this reason that even patients who were hypoxic were told to walk the hallways, while receiving oxygen.

A major complication associated with pulmonary diseases such as cystic fibrosis is the development of a pneumothorax. The existence of a pneumothorax requires immediate medical care and this can prove to be a life threatening situation. Patients report (and doctors I spoke with confirmed) that a pneumothorax is often extremely painful as is the treatment which is customarily carried out. The treatment of a pneumothorax requires much skill. Usually a chest tube is inserted to help the trapped air in the lung escape. Patients are only given a local anesthesia. Stronger painkillers such as morphine are contraindicated since narcotics depress breathing reflexes and would adversely affect the pulmonary status of the patient. After the chest tube is inserted, the patient usually receives much needed rest. The tube which is inserted into the patient's chest is attached to a piece of equipment called a Pleuravac. This is usually placed on the side or the front of the patients bed. Virtually all patients with a pneumothorax must remain in bed. In some cases, usually only after a few days, are they permitted to get out of bed to use the bathroom. As the patients condition improves,

he/she may be allowed to take a short stroll in the corridors, or more likely, to be situated in the patients' lounge. However, the patient still has a chest tube in place which is attached to a rubber hose and the Pleuravac. The Pleuravac itself is very noticeable and it makes noise caused by respiration and air in the lungs. The Pleuravac reminds some people of a bubble up filter you might find attached to an aquarium. Since the Pleuravac and the rubber tubing attached to the patient is so visible, it is often the subject of attention. One patient told me that while she was sitting in the patient lounge for half an hour, at least fifteen people in the lounge asked her " What is that ?. She told me " I am tired of people asking me what it is ------ I can't wait until this thing is removed ! ."

Summary:

In this chapter we have examined the relevance of Parsons' sick role model, the Szasz- Hollender model, Freidson's typology, and drawing largely upon Goffman stigmatization, to the situation of patients with cystic fibrosis.

Although Parsons' model of the sick role does not

completely apply to the plight of CF patients, nevertheless it does bear some relevance. In Parson's model model, as is the case with regard to Freidson's typology (in the category of serious deviation in which the individual is not held responsible for his/her situation), the person who is ill is expected to secure help and cooperate in their treatment.

Szasz and Hollender draw attention to variations in the doctor - patient relationship. Based upon the typology they present, it would seem that patients with cystic fibrosis best exemplify the " mutual-participation" model. This is based largely on the fact that CF patients appear to have major responsibility in terms of providing care for themselves. This responsibility is strengthened by what appears to be relatively close association with their health care providers.

Goffman's insights and contribution to our understanding of the societal reaction to various forms of deviance including illness, provide the basis for furthering our understanding of stigmatization which ill persons in our society frequently encounter.

This chapter also draws considerable attention to

cystic fibrosis patients' own recognition and strategies for coping with their illness. Some of the people adopted interesting and ingenious strategies in an attempt to hide the visible symptoms associated with their illness.

Chapter 4: COPING WITH THE REALITY OF CF

Victims of cystic fibrosis must learn to successfully carry on with their lives in spite of their disease. They often employ various strategies or techniques which serve to diminish their everyday self-awareness and consciousness of the disease.

Cystic fibrosis patients as well as social scientists who have studied them {Waddell} have identified different mechanisms which they believe help CF CF patients cope with their illness. Waddell (1985) suggests that Goffman's concept of neutralization is of key importance in understanding how CF patients deal with their disease. According to Waddell, in cystic fibrosis the neutralization process operates in conjunction with "faith" and "hope". Waddell states:

> "Neutralization is the process whereby confrontation with the epidemiological question of an illness is deflected; the uncertainties that serve to check or inhibit faith and hope are rendered inoperative and the people within the therapeutic network of relationships are free to engage in their exchange without serious threat to currency. Neutralization allows patients to remove from their own consciousness and self-awareness, the fact that they are combating a

life threatening illness." 1

According to Waddell, with cystic fibrosis there are five significant uncertainties which undermine faith and hope. These are:

1. etiology - the cause of cystic fibrosis is unknown

2. genetics - cystic fibrosis cannot be diagnosed early in pregnancy nor can the carrier state be detected

3. treatment - many factors in the treatment of CF are controversial

4. societal reaction - the disease is often difficult to conceal and frequently stigmatizes the individual and his family

5. life expectancy- the life expectancy of a person with cystic fibrosis is shortened but is indeterminate, ranging from a few days to thirty-plus years 2.

According to Waddell, through neutralizing these five uncertainties, faith and hope are reaffirmed. These provide the building blocks which enable CF patients to deal affirmatively with their situation.

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1. Waddell, Charles, FAITH, HOPE, and LUCK, { Washington, D.C.: University Press of America, 1983, pp. 28 ff.}

2. Ibid, pages 28-30.

The neutralization process serves as a catalyst for initiating and instilling faith and hope in the CF patients by removing from their level of consciousness an awareness of the uncertainties surrounding CF and its treatment.

Some researchers and medical sociologists have illuminated different paradigms which they believe serve the function of maintaining patients' ability to cope. Fox {1959} undertook a two year study of patients whose common denominator was illness, human experimentation, uncertainty, suffering, and death. Although the patients Fox encountered were not CF patients, CF patients experience many of the same influences as those in Fox's sample.

The Problem of " Not Getting Better ":

In her work, Fox discusses what she terms the problem of "meaning" and the problem of "not getting better." The problem of "not getting better" is one that frequently surfaces in the minds of persons with chronic ailments, degenerative diseases, or life threatening illnesses. Fox provides the following examples to illustrate the concept of " not get-

ting better ":

> Patient: Many of us have diseases you can't fight against, or help with, or cure. We're up against a stone wall....."3
>
> Dr. E: I talked to Mr. Kaye for almost an hour last night but I don't think it did very much good. He just doesn't seem to realize there"s almost nothing we can do for him. The problem is to make him understand that there isn't one medicine, or any medicine, that will cure him.....4

As Fox writes, when a patient acknowledges the problem of "not getting better," he/she is likely to experience frustration, disappointment, and discouragement.

The cystic fibrosis patients I spoke with indicated similar sentiments:

> "Bob was in here {hospital} last June for I.V. [intravenous] antibiotic therapy. He was hospitalized for 14 days. Then, suddenly on a Friday night, he complained of sharp chest pain, he had a hard time breathing, and he had a fever of 102 degrees. We went to the ER (emergency room) and he was back in the hospital for another two weeks of IV therapy. Since then he hasn't been feeling well. He has been "in and out" of the hospital. He's losing weight and coughing more. It just doesn't seem like he's getting

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3. Fox, Renee, op cit., p.130

4. Ibid, page 131.

better. 5

The comments of Bob's wife were similar to sentiments expressed by other CF patients and members of their family. Many people spoke of repeated courses of intravenous antibiotics in a relatively short period of time.

> "I think that this has been a lousy summer for all of the patients with cystic fibrosis, It seems like all of the "regulars" have been back and forth (in the hospital) this summer. The nurses tell me that Dr. J has had more CF patients on this unit in the past three weeks than the usual. My son has been hospitalized three times since May and it is only July. He's had a terrible time trying to catch his breath. They have him on oxygen. We were going to travel to Disneyland on vacation in two weeks but, that is out of the question until he is doing better than this !" 6

> "Dr. G has me maintained on three antibiotics intravenously, q6h {every six hours}....The usual combination: Ticar, Tobra and Naficillin...Boy, that Naficillin burns like hell! I'm getting drainage four times daily and the usual other medications. Not until they find a cure, or at least more effective drugs . do I expect to do better .....But then, this is CF, and I don't expect miracles. I've seen many CF patients whom I knew well pass on.....I'm living on borrowed time."7

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5. Wife of a cystic fibrosis patient

6. Mother of a cystic fibrosis patient.

7. A twenty-eight year old male cystic fibrosis patient.

The comments made by this young man draw attention to the grim and ever recurring events that many cystic fibrosis patients have soon realized. While antibiotic therapy may provide some benefit to some patients, it is not a cure nor does it provide long lasting treatment. In the eyes of this young person, until there is a cure for cystic fibrosis, the treatment is essentially palliative care, and in his own words, "he is living on borrowed time."

The problem of " not getting better " not only concerns the patient's own condition, but that of the other people that he/she comes in contact with. Cystic fibrosis patients tend to be aware of the progress that other CF patients were making as well. When fellow CF patients were not doing well, this often had a negative effect on the person's own spirits and emotions. As one patient stated, "Maybe, it is because we realize, we could be in their shoes, experiencing the fate that they are. "

The problem of " not getting better " also has an effect on family members. It is they who must endure the stress and trauma that frequently accompany a

downhill course in a patient's health. One woman told me that, once her son starts coughing " more than usual" at night and his clothes start looking big on him because of a loss of weight, she " right away" knows that things will be going downhill for awhile.

To a lesser extent, nurses and other health care providers are adversely affected by a decline in a CF patient's status. This is especially true when the patient was well-known to the staff over a lengthy period of time. (This is discussed in greater detail in Chapter 5.)

Communication as a Coping Strategy:

One method that some patients employed to help them cope was interaction and conversation with other CF patients. Through such networks, they came to realize that many of the experiences and events they encountered also struck other people. Many CF patients have suffered the agony of being out of breath (hypoxia, dyspnea), the physical inability to engage in certain activities (dancing, running, even walking) and not feeling up to spending a leisurely day or evening out with friends. As one CF patient stated, " It felt reassuring to know

that I am not alone. There are other people who have experienced many of the same problems." Sometimes they offered solutions for dealing with such difficulties. It would appear that informal channels of communication between patients may have a positive effect on the morale and psychological well - being of some CF patients.

I observed some adolescent CF patients confer with other CF patients regarding what a particular medical test { such as an echocardiogram or pulmonary function test} is like -- does it hurt? This was done in conjunction with information given to the patient by members of the nursing staff and/or physicians. Perhaps the patients place more credibility on what they hear from other patients than on the word of medical personnel. This is understandable in light of the fact that few doctors have ever had an arterial blood gas drawn { a painful procedure}; yet many cystic fibrosis patients have had blood gas tests done periodically.

Larter {1981} suggests that adolescent cystic fibrosis patients should be encouraged to talk about their feelings. Larter notes that this can have a

positive effect on body image and self - esteem by allowing the patients the opportunity to verbalize their feelings and frustration. Larter states , " Early recognition of stresses can be instrumental in solving problems before they become increasingly complex." 8 An open channel of communication could conceivably provide an early indication of impending stress.

Other Strategies:

Some CF patients have tried to deflect self awareness about their health and the limitations it imposes by keeping themselves actively involved with their work or personal interests. The brother of a recently deceased CF patient illustrate this point:

> "She knew she wasn't going to last long, so she kept as busy as possible" 9

Pinkerton et al {1985} suggest that CF people who were able to work full time tended to cope better than

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8. Larter,N, "Cystic Fibrosis", American Journal of Nursing, March 1981, vol 81:3, pp. 527-532

9. From the obituary of a 23 year old female CF patient. NEWSDAY, 10-28-85, p.15

other patients. Many cystic fibrosis patients are active members of the labor force. The patients I encountered were mainmly in white collar jobs or in the professions { law, nmedicine, science}. Many of these persons held advanced degrees and were active writers with published works { books, articles}.

The Problem of " Meaning":

Fox (1959) notes that the subjects in her study attempted to ascertain, or at least question why they personally were victims of illness. Fox terms this the problem of "meaning." The patients I saw were not really visibly concerned with this issue. Patients who I spoke with rarely devoted much thought to the question of " Why me?--Why do I have cystic fibrosis?. " A few subjects did point out that CF is genetically inherited but, in almost every case they noted that none of their relatives, nor their children were stricken with the disease. However, two of the patients had siblings who also suffered from the disease.

A number of years ago I served as a hospital volunteer. During that time, I encountered a woman who was taking care of her critically ill child. The woman

expressed her religious belief that the Lord gives a sick child to parents whom he believes can properly love and care for the child. Another woman, herself a patient, said she felt that, although she wasn't born well in this life, her "reward" for withstanding and coping with the pain and agony of CF would be a pleasant existence in the next world.

We must always bear in mind that persons with CF make use of coping mechanisms on a daily basis, and not temporarily or intermittently. As is often the case with other catastrophic illnesses, there are daily routines which must be followed.

Daily Routines:

I. Everyday Rituals:

For patients with cystic fibrosis a typical day begins with the immediate need to attend to their illness. Many patients with pulmonary disease report that, early morning and late night are the worst parts of the day in terms of breathing difficulties. A number of patients told me that, when they first arise, they normally cough a great deal. Physiologically, this is largely due to the fact that mucolytic secretions are

stationary and " plugged " since the individual is immobile during sleep. 10 Therefore, upon arising in the morning most patients receive their " inhalations " and engage in a form of physical therapy known as " postural drainage" or " P.D.", for short. Inhalations refers to the inhaling of medications such as bronchodilators [Isuprel, Bronkosol, Alupent, Proventil], mucolytic agents [Mucomyst], and/or antibiotics [Polymixin B, Colymycin, Kanamycin, Tobramycin, Gentamicin, etc]. Inhalation of medication takes a variety of forms.

The vast majority of cystic fibrosis patients receive inhalation medications through a compressor nebulizer or by an ultrasonic nebulizer. Most patients receive a bronchodilator diluted with saline followed by inhalation of an antibiotic. However, there is considerable variation among patients. Some patients receive Mucomyst followed by an antibiotic; others are maintaintained on Mucomyst and Bronkosol mixed together, followed by the inhalation of an antibiotic. Other

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10 Many CF patients report nighttime as being the most troublesome in terms of difficulty with breathing. Physiologically, this is due to the fact that PO2 levels decrease gradually during the course of the day.

patients do not inhale Mucomyst but receive Proventil Inhalation solution mixed with Intal.

It usually takes 20-25 minutes to prepare and carry out out the morning inhalation ritual. Most of the inhalation medications are manufactured as " already mixed" products. Therefore, the patient need only measure his dose, add 2 cc saline and pour the solution into the nebulizer medication cup, turn on the switch and inhale. Unfortunately, most of the antibiotics inhaled by patients with cystic fibrosis, are manufactured for IM, or IV use. As a result, these products are not premixed and often are formulated in powder form which must be reconstituted. Patients must therefore, inject 2-3 cc of .9% sterile sodium chloride [saline] into the vial, and refrigerate the vial prior to use. [This can sometimes be time consuming. In addition, depending on the medication, some antibiotics must be used within a few days after being mixed or they spoil. This prevents patients from mixing many vials at once and storing them in the refrigerator.] Once the medication is placed in the nebulizer cup and the nebulizer machine is operating, the patient need only sit back and relax. Most patients receive the medic-

ation via an oral mouthpiece. In the case of young children, sometimes the medication is delivered via a facial venting mask, while they sit on their mother's lap and she reads to them. During inhalations, many older patients sit back and relax by reading or watching television.

Postural drainage follows inhalation therapy. Postural drainage is a form of physical therapy in which the patient is placed in a variety of positions against gravity [usually on a slantboard or table] and vibration, percussion and vigorous clubbing is applied to various segments of the patients chest and lungs. The primary objective of postural drainage is to enable the patient to secrete and expel sputum and mucus from their lungs. These secretions if allowed to accumulate become problematic in that they breed respiratory infection, pnuemonia, and can over time yield other serious lung complications such as pneumothorax. In cystic fibrosis severe and permanent lung damage culminating in early death can result if mucus is allowed to accumulate unhampered.

Postural drainage itself is not difficult to under-

take. It is however time consuming and the amount of time needed to properly perform drainage varies from one individual to the next. This is largely due to factors such as 1) patient toleration [for example, patients are usually placed in a position known as Trendelenburg. In this position the slant table or bed is lowered straight down and the patient's chest is vibrated. Due to the extent of pulmonary involvement, many patients cannot breathe comfortably while in Trendelenburg. As a result, some patients receive oxygen during this position; in some cases the position is skipped altogether] 2) patient discomfort [some patients prefer a " vigorous " drainage, that is, they prefer when the clubbing is " hard" rather than "softer" ; others prefer it vice versa, and 3) productivity of sputum- some patients are more productive than others, that is, they are able to bring up more sputum. Some patients do not bring up sputum during the procedure, but will sometimes expectorate shortly after drainage is complete. If a patient is coughing and brings up secretions during the p.d., it will, of course, take longer to complete the procedure. Most older patients with cystic fibrosis perform postural drainage on themselves. However, some patients have

family members assist them in the procedure. A number of patients do drainage themselves three times daily, and have a physical therapist professionally perform the procedure two or three times weekly. 11 Younger children usually receive postural drainage from their parents. Parents report that they frequently play music, sing songs, or watch television during the procedure. They claim that this increases the childs cooperation and compliance with the procedure.

The degree of discomfort felt by the patient varies. This is influenced by various factors including the force of the clapping by the person performing the drainage, the patients weight, muscle and fat distribution, etc. There are also machines which can be purchased that perform postural drainage. Machines that are currently on the market include the Strom Percussor, and the Puritan-Bennett Vibrator/Percussor. Some patients told me that they prefer the Strom percussor to human hands. Other patients indicated the opposite preference. Postural drainage is viewed by many phy-

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11 Most physical therapists charge a fee of $ 60-75 for postural drainage. Usually the session lasts 45-60 min.

sicians as an absolutely essential protocol for the treatment of cystic fibrosis.

2. Other Common Procedures:

In addition to inhalation therapy and postural drainage, patients with cystic fibrosis must comply with additional regimens. The use of medications such as antibiotics, vitamin and nutritional supplements, pancreatic enzymes, and other medications is widespread. Many of the patients I encountered noted that they were receiving a number of medications. Due to pancreatic insufficiency and poor metabolic absorption, the vast majority of CF patients require pancreatic supplementation. Pancrease and Cotazym are the most widely used pancreatic enzymes by CF patients. The number of capsules of Pancrease or Cotazym that a patient consumed varied from patient to patient. Most patients took 2-3 capsules with meals and one capsule with snacks. In addition to receiving pancreatic enzymes, many patients use Polycose, Sustacal HC, or Meritene. These products are nutritional supplements. Some CF patients have more digestive damage then pulmonary involvement; in other patients the opposite is true. In patients with more

digestive involvement it is not unusual for the patient to be receiving 5 capsules of Pancrease with each meal, 2 capsules with snacks, colace or bacid, and possibly a nutritional supplement. Due to the pancreatic damage, CF patients require greater than normal doses of vitamins. Specifically, individuals with cystic fibrosis need megadoses of the fat soluble vitamins, vitamins A, D, E. Vitamin K is also frequently prescribed especially when patients are receiving two or more antibiotics simultaneously, when hemoptysis is present.

The Concept of "Suddenness":

Unpredictability is associated with many illnesses. Cystic fibrosis, is one of these serious conditions that arise without any warning or telltale signs. I shall refer to such situations as involving " suddenness." Such " suddenness " presents itself in a variety of ways: It can refer to [1] the initial phase in which a patient or members of his/her family learn that someone has been diagnosed as having the disease; [2] it can refer to the impact and adjustment process in the case of the patient has just learned he/she has cystic

fibrosis, or [3] it can also refer to medical emergencies and complications associated with the disease that arises spontaneously and may take the patient " totally by surprise ". I shall discuss each of these kinds of " suddenness".

Suddenness in the Diagnosis of CF:

As discussed in Chapter 1, cystic fibrosis is a disease which is present at birth. Today, many hospitals routinely test newborns for cystic fibrosis by means of the sweat test which is performed a few days after birth. This practice has emerged only within the past ten to fifteen years in most hospitals in the United States. Since this practice was not in effect earlier, many persons who have cystic fibrosis were not diagnosed during the first few days of life, but sometime thereafter. Conversations with some CF patients, seemed to suggest that there is much variance in terms of when patients were first diagnosed as having the disease. While many patients noted that the initial diagnosis of cystic fibrosis was concluded during early childhood, there were other patients who were not diagnosed as having CF until they were older. I met three

]people who first learned that they had the disease when they were young adults. When patients were diagnosed as having the disease in the later years, usually it was the gastrointestinal problems that seemed more troublesome at the time of the diagnosis. These patients also reported having fewer pulmonary complications or respiratory infections than patients who were diagnosed earlier in their life. All three of these people stated that the diagnosis came as a surpise. One person described it as " a shock ". She stated, " In fact we had never heard of cystic fibrosis... We didn't know what it was ". Thus, for some, the initial diagnosis of cystic fibrosis involves what I refer to as "suddenness."

In the course of doing fieldwork, I generally found that most people were fairly " open" and willing to discuss their lives and experiences in light of their illness. However, I noticed that many patients felt uncomfortable about discussing the events and circumstances which resulted in their being diagnosed as having the disease. Remarks such as, " That was back then, I hardly remember" or " Things are different now " reinforced this feeling. In undertaking this endeavor, one of the things I wanted to prevent as much as pos-

sible was making people feel uncomfortable or exploiting people whom I encountered. To this end, I felt it would be inappropriate to query people as to the origins of their diagnosis, since this seemed to be an unwelcome area to probe.

Although I chose to address this aspect of cystic fibrosis superficially, there was one person, an adolescent female, who did not seem to mind telling me how, at the age of 17, she learned that she had cystic fibrosis. The events leading to the discovery that she has CF are most interesting: She had been brought to the hospital after complaining of pain in her abdomen. She was diagnosed as having appendicitis and was operated on to remove her appendix. After the surgery she was feeling much better. During the course of conducting routine rounds, a resident in pediatrics noted that she was coughing and had a dry hacking cough. When the physician asked her about the cough, she told him that she often coughed and attributed it to a possible allergy. Apparently, the doctor chose to investigate further. He discussed the matter with her private physician. A few days later, she went for a sweat test,x-rays, and a pulmonary function test. She

was then diagnosed as having cystic fibrosis. It is interesting to note that it was the interest of a young, relatively inexperienced, pediatrician in the nature of a patients cough that led to the diagnosis of cystic fibrosis. This situation illustrates the concept of " suddenness " in that a person presumed to be healthy undergoes an appendectomy { which is a fairly common procedure } and is suddenly and unexpectedly found to suffer from a potentially life threatening illness. In the words of this patient, " I expected to have my appendix removed and to go home. I never dreamed that they {doctors} would find anything wrong with me ."

2. Suddenness Following a Diagnosis of CF:

The concept of "suddenness" also applies to the status of the newly diagnosed patient who soon learns the " ins and outs " of medical treatment for the disease. Almost immediately following diagnosis, patients are provided with information about cystic fibrosis, and the medical protocol that they personally will have to follow.

It is reasonable to expect that a patient who has been diagnosed at birth as having the disease , is not

as likely to experience as much grief and shock as a person who learns later in life that he/she is ill. The person who is diagnosed as having CF at birth, has probably adjusted and accepted his/her situation in a short period of time; one would not expect that the daily routines and the fundamentals of care for CF come as a surprise to a person who has been treated for the disease for a long period of time. For example, the patient who has undergone treatment since childhood knows that he/she requires pancreatic enzymes with each meal and possibly with snacks. He/she is not likely to forget to take their enzymes with their meals, or to neglect other aspects of their medical care. On the other hand, the situation of the newly diagnosed patient [especially if the patient is diagnosed as having CF later in life] is very different. The patient who is assumed to be healthy and who carries out a " normal lifestyle " is likely to experience grief, uncertainty, a nd a sense of " suddenness." For example, if we refer back to the situation involving the 17 year old girl who had an appendectomy and suddenly learned that she has cystic fibrosis, the diagnosis is likely to be met with denial, resistance, and shock. The adjustment process

for such an individual is profound. This individal will probably have to change certain aspects of her lifestyle to accommodate the accepted " rituals" [such as taking Pancrease or Viokase] with each meal, and doing postural drainage three times daily. In essence the adjustment process for this young woman represents a kind of " rite of passage." This individual's status changes from that of a " healthy young woman " to that of a " person who is ill and requires medical care and attention."

3. Suddenness and Emergency Situations:

A medical emergency can befall any human being, whether they are healthy or under medical treatment for an illness. In cystic fibrosis, changes in a patients condition are often marked by sudden and unforeseen complications that arise. Emergency situations that are common for some patients with cystic fibrosis include increased cough accompanied by shortness of breath {dyspnea], coughing up blood or the streaking of blood in sputum [hemoptysis], sharp or intense chest pain , fever accompanied by a feeling of malaise. All of these symptoms may be indicative of serious illnesses such as pneumonia, bronchiectasis, atelactasis, a haemophalus or

pseudomonas infection in the chest, or perhaps, most serious of all of these conditions, a pneumothorax.

Based upon discussions I had with some of the patients it was interesting to discover that most emergency situations they encountered tended to occur during the late evening [between 9 PM - midnight] or on weekends. Friday nights seemed to be popular in terms of the number of patients who suddenly took ill and who were admitted to the hospital { after being evaluated in the emergency room } for treatment. The CF center at the hospital was undoubtedly aware of this trend, since it was their practice to have at least one pediatric pulmonary fellow and an attending physician " on call " for emergencies at all times. Two resident physicians I spoke with suggested that the pulmonary fellows and attending physicians " on call " for CF, anticipate that in all probability they will be summoned back to the hospital to see at least one patient in the ER per night One of the doctors told me that on one holiday weekend, four CF patients took ill and had to be evaluated in the ER. All four required hospitalization and he remarked " Although it was the doctor's wedding anniversary she returned to the hospital and spent almost six hours

examining the patients, waiting and tracking down lab results, and then writing up admissions orders ..... By the time she arrived home, it was probably time for her to return for morning rounds ."

Apparently, the inopportune timing of medical emergencies was not limited solely to physicians. One patient told of an " unexpected" situation that she experienced:

> " I was busy baking a cake when I felt a pulling sensation in my upper arm and a sharp pain in my chest. Then I started spitting up blood. I was in such pain, that I could hardly move. My parents called 911 and they took me to the emergency room. Sure, enough...I had another pneumothorax . I was planning to go to a party that night.

This patient's assessment of a recent episode draws attention to the " suddenness" associated with the onset of illness.

Interestingly, the " suddenness" and " unanticipation " of a medical emergency, are not confined to the outside world, as the following example illustrates:

> " After being in the hospital for two weeks on I.V. therapy, I was really looking forward to going home. I was watching Concentration on TV, when I felt a sticking pain

> in my chest. The pain seemed to be getting worse, so I told my nurse. Erik (the doctor) came in and he took a listen to my chest. He said it didn't sound too bad, but he wanted to get another X-ray. After they took the X - ray, I was in the lounge playing Foosball. I was feeling a little better. A short time later, Gail [nurse] came in, and said " I've got bad news... guess what kid? You've got a pneumo. "

This patient wanted very much to leave the hospital and return home. Perhaps, the furtherest thing on her mind was that after two weeks of hospitalization, she would suffer from a pneumothorax. This situation is unusual in that most CF patients who have undergone I.V. antibiotic therapy for two weeks, would not ordinarily expect a pneumothorax to follow. This situation is important because it draws attention to the " suddenness" and unpredictability which accompany serious and life threatening illnesses such as cystic fibrosis.

Further Risks and Dangers:

In addition to dealing with the problems of suddenness and medical emrgencies, it is recognized by the patients themselves that there are major risks associated with their treatment. A source of danger, and perhaps the most important one, is the medications used

to combat the disease. As discussed previously in this chapter, patients with cystic fibrosis consume a wide variety of medications including pancreatic enzymes, nutritional supplements, bronchodilators, oxygen, mucolytic agents, and antibiotics { intravenously as well as orally }. In addition, since many patients have other diseases secondary to cystic fibrosis, they may be receiving additional drugs such as steroids, blood pressure or diabetes medicines.

Although medications can be useful, and perhaps life saving tools in fighting disease and prolonging life, they nevertheless carry along with their use certain risks. Some medications have a greater margin of safety than others. For example, it is believed that the use of pancreatic enzymes such as Pancrease and Cotazym in cystic fibrosis is not only essential but also posses little risk of side effects to the patient. The only known contraindications to pancreatic enzymes { such as Pancrease, Cotazym, or Viokase } is in patients allergic to pork or rarely, a patient who is receiving extremely high doses of these enzymes may have high levels of the by products of the enzymes " spill over " into his/her urine.

Unfortunately, many of the other medications routinely used in the management of cystic fibrosis are known to demonstrate side effects and dangers associated with their use. A case in point concerns the antibiotic, Chloromycetin { Chloramphenicol }. Chloromycetin is often used in cystic fibrosis, especially when a patient possesses organisms susceptible to the drug, and the patient is not responding to less harmful antibiotic agents.12, 13 The Physicians' Desk Reference { PDR }, recognized by physicians as the " authoritative source " on pharmaceutical agents, has a **WARNING box** which contains the following information about Chloromycetin:

> " Serious and fatal blood dyscrasias (aplastic anemia, hypoplastic anemia, thrombocytopenia, and granulocytopenia) are known to occur after the administration of chloramphenicol. In addition, there have been reports of aplastic anemia attributed to chloramphenicol which later termin-

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12 The choice of which antibiotic(s) are prescribed is based on the results of sputum cultures. From sputum, organisms are grown in the lab and then tested for sensitivity to various antibiotics.

13 In CF, the following organisms are frequently isolated from sputum: Pseudomonas auerginosa, Staph. aureus, E.coli, Klebsiella, and H. Influenza. This information was derived from a conversation with a physician.

> ated in leukemia. Blood dyscrasias have occurred after both short term and prolonged therapy with this drug. Chloramphenicol must not be used when less potentially dangerous agents will be effective.....it must not be used in the treatment of trivial infections or where it is not indicated, as in colds, influenza, infections of the throat, or as a prophylactic agent to prevent bacterial infections.
>
> Precautions: It is essential that adequate blood studies be made during treatment with the drug. While blood studies may detect early peripheral blood changes, such as leukopenia, reticulocytopenia, or granulocytopenia, before they become irreversible, such studies cannot be relied on to detect bone marrow depression prior to development of aplastic anemia. To facilitate appropriate studies and observations during therapy, it is desireable that patients be hospitalized. 14,15

In the clinical setting, physicians openly and readily acknowledge the possible risks associated with the use of Chloromycetin, and the imperative need to closely monitor and follow patients receiving the drug. Some cystic fibrosis patients are permitted to receive the antibiotic on an outpatient basis, but they are required to report back to the hospital every 2-3 days for blood work { a CBC, with retics, and platelets, }. At a meeting of CF patients, one phys-

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14 Physicians' Desk Reference, 42nd edition, under Chloromycetin- Parke Davis.

15 It should be noted that the PDR lists under the Indications: cystic fibrosis regimens.

ician of the CF team told the gathering:

> " Chloro is a good drug but you've got to watch it like a hawk. Not only do I have to have blood work done every few days, but other studies are needed as well. I try to keep patients on it a for as short a time as is possible, preferably for 10 days to two weeks at a time. But, it can be used in CF for longer periods of time, if the blood work is fine-- there is no evidence of the Chloro effect. Incidentally, after a week on chloro periodic eye exams are needed to make sure there are no eye complications { you can get an optic neuritis from it } and you need to be seen weekly to make sure you don't show signs of a peripheral neuritis, that is pain in the feet".16

The dangers and risks associated with medications employed in the management of cystic fibrosis is not limited to the use of Chloromycetin. Other intravenous antibiotics sometimes used in CF include Naficillin, Vancomycin, and, Amikin. Patients receiving these medications must be closely monitored and observed since serious side effects are known to occur with their use. Side effects experienced by some patients include marked nausea, diarrhea, intense pain and phlebetes at the IV site, as well as endocrine and gynecological problems.

Patients I have spoken with told of situations in

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16 These remarks were made at a meeting which was attended by patients and physicians alike.

which they had to sign" Informed Consent " and other hospital forms because they were not adequately responding to therapy, and it was strongly advised by the CF physicians that they undergo a trial run on " experimental drugs " { ie, antibiotics that were about to be released pending FDA approval }. Cystic fibrosis patients are usually given access to antibiotic agents { especially anti Pseudomonal agents } prior to FDA approval and release for distribution to the general public. An example, is the recently released antibiotic Cipro { ciprofloxacin }. One patient who agreed to try the drug when it was under experimental study had to be taken off of it because he was developing side effects. Patients who agree to take experimental drugs are taking risks in that the medicines may prove to be beneficial but, there is the ever present possibility that serious and perhaps, even fatal side effects can ensue. The willingness of some cystic fibrosis to try experimental drugs stands as a testament to the degree of seriousness and urgency associated with their disease.

Summary:

This chapter has examined coping mechanisms and the everyday realities of living with cystic fibrosis.

Coping with cystic fibrosis involves two important facets: {1} the individual must learn to accept his or her illness, and {2} the individual must learn the routines and daily rituals for treating their illness.

Acceptance that a person has cystic fibrosis can be accomplished in a variety of ways. Waddell suggests that cystic fibrosis patients on one level acknowledge the particulars of their situation, but they also " neutralize" that awareness by placing it below their immediate consciousness in the course of their everyday lives. Larter suggests that communication among cystic fibrosis is most beneficial as a coping mechanism, since it allows them to verbalize and share their experiences with others who have the same plight. My research reveals that some patients do in fact, as Waddell suggests , appear to "neutralize" various aspects of their disease. However, I also found that this was not always the case. The anectdotes and remarks made by some of the patients suggest quite the opposite... namely, that some are aware of the grim realities surrounding cystic fibrosis, and they cannot simply place them below their level of consciousness. The fact that there were strong networks of communication and long standing friendships

among various cystic fibrosis patients [especially adolescent females] suggest that Larter's observation that communication is an integral coping mechanism is most accurate.

In addition to recognizing ones status, successfully dealing with an illness mandates that a person properly care for his/her own medical needs. As discussed in this chapter, cystic fibrosis patients must learn to perform many time consuming, and perhaps unpleasant procedures such as postural drainage, inhalation therapy, adherence to dietary restrictions, and consumption of massive medications { orally, inhaled, and injected } on a daily basis in order to survive. This regimen goes on even while the patients are being followed closely by physicians. Regular visits augmented by the repetition of diagnostic and evaluative tests { spirometrical studies, lab and blood work, periodic X-ray studies, and painful arterial blood gases } are all common attributes of medical treatment of cystic fibrosis. The patient who wants a chance to survive must diligently abide by the prescribed treatment plan for his/her predicament.

The latter part of this chapter examines the impact of medical situations that arise suddenly and without warning. In cystic fibrosis, it is interesting to note that the initial diagnosis of the disease is often made spontaneously and sometimes, under pecular circumstances { such as the girl who after undergoing an appendectomy learned that she had cystic fibrosis because a doctor found her cough to be pecular }. The concept of "suddenness" also applies to the newly diagnosed patient who is suddenly thrusts into a situation where he/she must transform a lifestyle from that of a "healthy person" who has no medical restrictions placed upon them, to that of a " ill person" who must take special steps and precautions to safeguard their health. In addition to both of these situations, we find that "suddenness" occurs in connection with the onset of medical emergencies that victimize some patients with cystic fibrosis. Conversations with patients disclosed that many medical complications and emergencies occurred at the most inopportune times, were totally unforeseen, and beyond the individual's own control. Patients in my sample seemed to indicate that medical emergencies are most likely to occur from 9 PM until 12 midnight.

Furthermore, the mystique that accompanies the onset of illness is not soley limited to time, but also to place. Generally, physicians would not expect a CF patient who has made satisfactory progress and who has just received megadoses of I.V. antibiotics to succomb to a pneumothorax.

This chapter also draws attention to the dangers that are forever present concerning the use of medicines which are commonly prescribed in treating cystic fibrosis. Patients must not only contend with the possibility of side effects emanating from drugs that are widely used by the general public, but they may also experience serious side effects from experimental drugs { drugs that for the most part that have undergone limited human and clinical trials }.

Chapter 5: THE HOSPITAL SCENE

Background:

In his monumental work, The Care of Strangers, Rosenberg presents the historical development of the hospital in American society. The role of hospitals and the functions they serve has changed dramatically over time. In 1800, hospitals were an insignificant aspect of medical care, and no gentleman of property or standing would have found himself in a hospital unless stricken with insanity or befelled by epidemic or accident in a strange city. 1 Slightly over a hundred years later, in 1909, a census of American hospitals located 4,359 with 421,065 beds { a total that did not include mental or chronic disease hospitals such as tuberculosis sanitariums }. 2 Not only had hospitals increased sharply in number, but individuals of all social class standings began to avail themselves of their services. 3

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1 Rosenberg, Charles, The Care of Strangers: The Rise of America's Hospital System, { New York: Basic Books, 1987, p.4 }

2 E.H.L. Corwin, The American Hospital, The Commonwealth Fund, New York, 1946, p.8

3 Ibid

Although the popularity of hospitals grew dramatically since the 1800's, many of the services and kinds of care associated with hospitals today were previously met by an individual's own family. For example, at the turn of the century, women usually gave birth at home. They were tended to by family members, neighbors, or perhaps, in some cases, midwives. In contrast, today most American women give birth in hospitals and are tended to by physicians, obstetricians or other medical personnel. Similarly, in previous times, chronically ill persons were often tended to by the members of their immediate family. As Rosenberg notes, " to most observers the twentieth century hospital seems an inevitable, if perhaps imperfect, institution, one that grew unavoidably out of the interaction between social necessity and an emerging technical capacity. 4

Hospitalization in Cystic Fibrosis:

There are limits to medical care being provided on the homefront. While " everyday rituals " such as postural drainage, inhalation therapy, and the taking

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4 Rosenberg, op cit., p. 10

of oral medications are easily carried out at home, from time to time more aggressive treatment needs to be implemented. For example, if a patient is losing ground in terms of his/her pulmonary status, hospitalization for intravenous antibiotic therapy with multiple antibiotics is a distinct possibility. While intravenuous antibiotic therapy can be accomplished at home for patients using one intravenuous antibiotic , when patients require two or three antibiotics simultaneously, hospitalization is considered the universal norm among physicians. Another situation which calls for hospitalization is Total Parenteral Administration , or TPN for short. TPN is administered to patients who are either unable to eat, or who fail to consume enough caloric intake necessary to survive. In TPN, patients are fed huge amounts of carbohydrates and essential minerals and supplements directly into their bloodstream. Patients who are receiving TPN must be closely monitored. During serious illness or following major surgical procedures, cystic fibrosis patients are sometimes placed on TPN.

TPN represents one condition which would require a patient with cystic fibrosis to be hospitalized. There

are other circumstances which also result in patients requiring hospital care. As noted in Chapter 1, the CF patients I studied, were treated at a CF Center located within a Children's Hospital. The CF Center follows patients of all age groups, from the first few days of life into adulthood. Although the emphasis is on providing patient care on an outpatient basis, cystic fibrosis patients like other seriously ill persons require hospitalization on an ongoing basis. At any given time about 1-3% of the patients enrolled in the CF Center are hospitalized. 5 This figure suggests that of the approximately 200 patients in this center, at any given moment 2 to 6 patients are " inhouse."

Conversations with CF patients suggest that there is much variation among patients in terms of how often they are hospitalized, and how long their " usual " length of stay lasts. [For example, I encountered one patient who requires hospitalization an average of once a year and he usually remains in the hospital for slightly over a week. This appears to be considerably

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5 This figure was provided by physicians at the CF Center, where the patients I encountered were receiving care. It should be viewed as a general " ballpark" figure

less than the norm in comparison to other patients of the same age.] 6 Based on conversations with adults with cystic fibrosis, it seems as though most patients require hospitalization about two or three times per year. Their usual varies runs from five to fourteen days per hospitalization. Many of the patients told me that their hospitalizations do not create problems with their family life or in terms of their employment. One strategy used by some of the patients was to try to schedule hospitalizations when they were on vacations, or during the lull or slow periods at work. Similarly, college students or those who were on an academic schedule tried to come into the hospital during holiday recesses { spring, winter, fall, summer, etc.}

Patients who spent five days in the hospital usually represented one of the following situations: {1} they were going to receive I.V. antibiotic therapy at home after their discharge. Such patients are hospitalized to begin their I.V. therapy, and to establish the proper dose and " safe levels " of the

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6 Although this patient does not represent the "model" or "average" patient, he illustrates the diversity that occurs regarding the length of hospitalization required by CF patients.

antibiotic. 7 {2} the patient was hospitalized following a brief illness such as a viral infection or flu, {3} other illnesses unrelated to cystic fibrosis. For example, one patient underwent a minor surgical procedure { a tonsillectomy }. Her surgeon chose to keep her in the hospital a day longer than usual, because she has CF. It is important to realize that each patient's condition is unique.

Some patients respond more readily to antibiotic therapy than others. Some patients show considerable clinical improvement within 24-48 hours after receiving their first dose of antibiotics, while for other patients recuperation and improvement are slow. Unfortunately, there are patients who show minimal improvement, or who require " trial runs " with different antibiotics and medications, before a combination is discovered which provides measurable benefit to the patient. There are also situations in which the patient is responding to the antibiotics but, due to

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7 This is especially the case with respect to the aminoglycoside antibiotics such as Tobramycin, Kanamycin, and Gentamicin. These agents are known to cause nephrotoxicity, and ototoxicity.

complications, the dosages of the antibiotics had to be lowered or discontinued. This often happened when the patient was suffering from diabetes mellitus, kidney or hepatic impairment secondary to cystic fibrosis. All of these situations and factors can and often do play a major role in the determination of how long a cystic fibrosis patient needs hospitalization.

Since the CF Center functions under the auspices of the Children's Hospital, it is standard hospital policy that all CF patients { regardless of age } be treated by the Pediatric Pulmonary - Critical Care team. Hospitalized CF patients are found throughout the entire hospital. Patients who were 12 years old and above { including adults } were ordinarily assigned to the adolescent unit. 8 Adult patients who require intensive care are placed in Pediatric Intensive Care. A few adult patients told me that they didn't mind being treated in Pediatric Intensive Care when they

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8 Although some of the older patients were not particularly pleased with this arrangement, they had to learn to endure it. Physicians tried to instill in patients the idea that the nurses on the adolescent unit were more atuned to the needs of CF care, than nurses on the adult floors -- where they seldom dealt with cystic fibrosis patients.

were seriously ill because they found the staff to be well informed about their disease. Patients seemed to reflect more positively on the quality of the care provided in Pediatric Intensive Care, than on the adolescent unit.

Since all of the CF patients I was introduced to were "regulars" on the adolescent unit, in order to understand what hospitalization entails for CF patients, it is necessary to outline some details about the adolescent unit.

Patients' Rooms:

The adolescent unit consists of fourteen patient rooms, most of which accommodate two patients. Each room has a private bathroom with a shower stall, a large sink with a paper towel dispenser, two small lockers designed to hold a patients suitcase, a small closet { some rooms }, two or three rollable metal carts { each containing a bedpan, steel bath basin, urinals, sputum collection cups, urinalysis cups, alcohol swabs, surgical tapes, and a box of hospital tissues }. Hanging from the center of each room are two

color television sets with remote control tuners.9 Hanging from the ceiling are moveable IV holders. Each room is equipped with beds capable of being transported. The beds are not like those most people are accustomed to observing in a hospital setting. On the contrary, the beds in this unit represent the latest in modern electronic technology. Each bed can be positioned to the patient's own personal satisfaction. At the mere touch of a button, a patient can lower or raise the height of the bed, the position of the bed, as well as the angle of the bed near the patient's feet. The beds are the most comfortable one can imagine. For the few patients who are bedridden, anxiety is greatly minimized because of the features of the beds. Above every bed there is a two - way intercom system, as well as a pushbutton device which permits patients to summon the nurse when necessary. One of the unique features of the intercom system was the ability of the unit receptionist to simply press one button on her unit, and to announce her message throughout all of the rooms. This is quite useful in

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9 Television sets are for rent at $4 per day Two color televisions are also in the patient lounge for all persons to use.

situations such as trying to locate a nurse who might be attending to a patient. All the receptionist need do is push the button and state, " Carol, check the IV in room ___ , and regardless of where Carol was she could receive the message and respond accordingly. Each room also had its own heat and air conditioning unit, which was under the patient's own control. Next to each of the beds was a flowmeter, and separate wall outlets ready to disperse either air or oxygen.

Patient Lounge:

In the middle of the hallway on the adolescent unit is the Patient Lounge. The room is also referred to as the " rec room" by younger patients. On the walls pleasant posters featuring vacation spots or cartoon characters such as Mickey Mouse and Snoopy are displayed. Attractive furniture highlights the room. There are two color televisions, a stereo, and a computer video arcade for patient use. There is also a bumper pool table, and foosball. 10

Kitchen:

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10 This is not a misspelling for football. Foosball is a game similar to table top hockey or nok hockey.

The adolescent unit has its own kitchen for patient use. Patients may store in the refrigerator any perishable items they have. The refrigerator is full size { 21 cubic feet } and is well stocked with jello, custards, juices, milk, and other foods. There are foods for persons on " regular diets " as well as those on special diets. The kitchen also has a coffee pot { for $1.00 a week patients can participate in the coffee pool which allows them unlimited coffee }. There is also a full size stove available for patient use. Sometimes patients will save a portion of their meals, or wish to heat up their snacks at a later time. The stove is ideal for such occasions.

The Snack Cart:

Everyday a snack cart makes its way around the halls of the adolescent unit, maneuvered by a hospital volunteer. The cart usually arrives on the unit around 2-3 P.M. The cart is usually well stocked with juices, milk shakes, frozen fruit bars, Bungalow Bar sundaes { strawberry and chocolate },ice milk, jello { regular and diet }, small packets of cookies, and bags of pretzels. Patients whose medical status permits. are allowed access to the snack cart.

Daily Walks:

Realizing that hospital inpatients sometimes experience the " hospital blues " at least once a day patients are allowed to leave the floor escorted by a nurse or a nurses aide, to visit the gift shop, or to sit on the outside grounds of the hospital. Daily walks were intended to alleviate the dreariness of being a hospitalized patient. This activity was in addition to occupational or physical therapy which some patients were receiving.

Special Events:

A few patients remarked to me that the best time to be hospitalized is around the holidays. During major holidays, there is almost always a special event or party in the works. For example, during Christmas, Santa Claus makes his annual visit, Ronald MacDonald pays a surprise visit , and sometimes, various food concerns donate food and/or candy for hospitalized patients and staff.

Clothing and Attire:

In many hospitals, there is a formal dress code for staff as well as for patients. This is not the

case on the adolescent unit. Nurses as well as doctors seldom wore white uniforms or other attire associated with the medical profession. Most nurses wore colorful pantsuits or blouses. Doctors tended to wear street clothing.

The policy of the hospital is to allow patients wishing to wear their own clothing to do so as long as it was not injurious to their health.11 I observed no relationship between the nature of a patients illness, the severity of a patients condition, and the likelihood that he/she would wear either pajamas or regular clothing. A number of patients stated that they felt more ar ease wearing street clothes as opposed to thpajamas when they are hospitalized.

A Hospital None the Less:

Although the adolescent unit possesses many things not typically associated with hospitalization { the daily walks, the snack cart, the holiday parties, patient access to a kitchen }, one should not get the

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11 Most patients were able to wear street clothing. Even patients who were receiving I.V. meds are able to wear a shirt or blouse. This is possible as a result of the use of heparin locks.

impression that " life on the unit " is all fun and games. As Slaby and Glicksman [1985], two physicians, emphasize " hospitalization causes stresses in addition to those of life threatening illnesses ... locus of treatment determines, to some degree, how a patient copes with treatment." 12 Behind this facade, there is important and perhaps life prolonging medical care that is being provided to patients with a life threatening disease. Perhaps all of these elements are necessary to minimize and alleviate patient stress and the " hospital blues " during hospitalization.

Goffman [1961] in his work, Asylums, discusses the concept of " total institutions ". Total institutions refer to institutions in which the individual is under the control of other people who are in a position of power and authority. Goffman's concept of total institutions applies to such places as prisons, schools, mental asylums, and hospitals. Although there exists some degree of choice and patient autonomy on the adolescent unit, one should not lose sight of the

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12 Slaby, Andrew E, M.D., and Glicksman, Arvin, M.D., Adapting to Life - Threatening Illness, { New York: Praeger Pub, 1985, p. 220 }

fact that the unit is part of a hospital which is a bureaucratic structure, founded upon rules and regulations. Since patients are not permitted complete freedom to do as they please, and there are regularly scheduled activities { daily walks, patient activities, etc. } the adolescent unit can be seen as possessing some characteristics in common with other total institutions.

Depersonalization:

One of the consequences of hospitalization is the depersonalization that follows. Hospitalized patients give up privacy and aspects of their own identity. For example, most rooms on the adolescent unit are semi-private rooms intended for two patients. Hence, patients forfeit some of their privacy by having to share quarters with another individual ..... one with whom they are unfamiliar.

Rosenberg elaborates upon this depersonalization in connection with the hospital admissions procedure at the turn of the century. Rosenberg states:

> " By the end of the century, admission had become a routinized, physically segregated procedure. A standard physical examination, preceded ad-

> mission to a bed, as did a compulsory bath and, in many institutions, delousing. The patient's street clothes were removed, locked away, and replaced with a hospital gown..... The admissions process might be thought of as a ritual separating the patient from his or her previous identity --- and especially those class specific attitudes, behaviors and possessions administrators sought to exclude from the wards. 13

" Regulars ":

Since CF patients from age 12 until adulthood are hospitalized in the adolescent unit, most of the patients soon become " regulars ". I shall use the term " regulars " to refer to those patients who are readily recognized by the nurses and fellow patients on the adolescent unit, as a result of being repeatedly hospitalized.

The " Home Away From Home " Syndrome:

A person who is hospitalized for the first time is likely to feel lonely, alienated, and perhaps insecure. He/she does not know the " ropes " of being a hospital patient. The " newcomer " to the hospital setting is unfamiliar with the proper etiquette of hospital behavior since he/she is not necessarily aware of the

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13 Rosenberg, ibid, p. 292

norms governing patients' obligations and responsibilities. As a patient is hospitalized repeatedly within a relatively short period of time, there is a possibility that he/she can become very or overly secure with his status as a hospitalized patient.

The " home away from home " phenomenon can be said to have occurred when: {1} a patient comes to regard the hospital as a security blanket { a place where he/she finds protection from the outside world } or as a " second home " { in the same sense that a person who is semiretired comes to regard his/her vacation home, {2} a patient feels that he/she is in no rush to return home or, may in fact be disappointed that they are about to be discharged. Perhaps, for some people the " home away from home " phenomenon functions as a form of escapism.

I observed a number of teenage patients who seemed very content with the role of a hospitalized patient. One patient told me, " I love it here... the food is great, most of the nurses are nice, the televisions are super, the activities in the day room are fun, and I get gifts from my friends". The only bad

thing is that I've have to be in here for IV medicines and I don't like being stuck { angio catheter}. Judging from the way this person reflected upon the hospital, it sounded more like a resort or hotel.

It is interesting to note that the " home away from home " phenomenon is not limited to teenagers. I observed an older patient saying openly to a nurse:

> " I'm really in no rush to go home. Are you sure that I am really ready to go home ? I've only been in here about eight days, which isn't very long."

I was surprised to learn that this patient was a married man with a wife and two children, and that he was on vacation at the time that his illness struck.

Patient-Staff Relations:

Most patients seemed to like the nurses who staffed the adolescent unit. Similarly, nurses whom I spoke with seemed to feel comfortable and content in their relations with cystic fibrosis patients. One nurse remarked, " I know so many of those patients from over the years that I know them almost as if they were members of my family ." In fact, after conversing with some of the patients I had the feeling that some

of the patients looked upon some of the nurses as members of their " extended family." While sitting in the lounge, one patient was overheard telling another patient:

> " Alot has happened since you were last here. I remember you were here last summer. June left to have her second baby. She had a boy. Sue is on vacation now... she went to England. Bonnie is on vacation. Donna Marie left to work somewhere else. Steve doesn't mop the floors anymore...he hurt his knee....some other guy {sic} does it now. Janet { the dietitian } left, and the new one is a real bitch. 14

It is interesting to note how interested and aware patients are of the personal lives of members of the staff. The opposite also appeared to be the case. Nurses on the adolescent also seemed to keep " tabs " on the latest news and developments concerning patients whom they cared for on a long term basis. Nurses would sometimes ask one CF patient if they had seen or heard anything about another patient. One nurse's aide who I know since my days as a hospital volunteer remarked, " Let's face it love..We're all one big

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14 All of these names are psuedonyms for real persons.

happy family." Although this person's sentiments might seem heartwarming, they appear to be slightly optimistic.

Beautiful People:

There were a few members of the staff who really tried to make the hospital setting as comfortable and as pleasant as possible for patients. Bill { pseudonym }, the youth-life specialist { discussed in Chapter 1 } was one such person. He tried, and was quite successful, in engaging mobile and bedridden patients in activities, computer video games, and crafts which helped pass the time of day.

Another truly devoted person was Tenaj { pseudoonym }, the dietitian. She once told me and another member of the staff, " I want to do all that I can to make the patients feel as happy and as comfortable as possible. Having to be hospitalized is bad enough for anybody; it is especially depressing for a child or a young person, or if you have to be hospitalized regularly." Tenaj did all that she possibly could. Unfortunately, in many serious illnesses such as cystic fibrosis, when patients are ill they may lose their

appetite. Furthermore, CF patients must consume a diet that is high in caloric intake due to their pancreatic insufficiency. Tenaj frequently bombarded CF patients with enormous quantities of food. She handed each CF patient, anorexic, or other critically ill patient on the unit, a list of all of the foods, pies, snacks, drinks, etc. that the hospital kitchen stocked. Patients were told to " write in and circle" the foods that they desired. When the daily menus were returned, she would initial the menus and write "OK", which was the cue for dietary workers to place all of the times on the patients trays. Tenaj's dedication was not only recognized and appreciated by the patients; the nurses and other staff on the adolescent unit also took notice. One dietary worker told me, " She's a real nice lady. I've been working at this hospital for over seven years and I ain't never seen {sic} any dietitian on any floor give us a hand... When we bring up the trays, if she's on the floor, she helps distribute the trays." Tenaj frequently appeared on the floor during meal times and she would go to each and every patient asking, " Can I get you anything else ? Are you sure ? .. If you want another portion or if

you don't like what you ordered just let me know, and I'll order you something else ..... I want everyone eating."

Adults in a Child's World:

Some of the older CF patients whom I spoke with indicated that they felt a little " out of place " or awkward being treated on a unit intended for adolescent patients. Two of the patients suggested that this feeling was perpetuated in large part by the attitude of the head nurse. One patient told me:

> " June [psuedonym] really resents the presence of older patients on this unit. She sees us as a threat to her authority and autonomy. I think she feels that without the older CF patients, she would have greater control and command of the floor. What she doesn't realize is that it is her job to see to it that we are properly treated".

One of the nurses told me that cystic fibrosis patients are usually easy to care for. For one thing, they know the routines and they generally comply and cooperate with us { ie, the nurses }. Furthermore, since they know all about I.V.'s, " They will keep you posted as to how their I.V. is doing. They know when the buretrol is running low and when they need to call

us for a flush, or to be put on a lock { heparin lock} ..... Some of the cystics even know how to reset the IMED { I.V. machine } and will let it run KVO if we're busy at the moment " . While visiting a CF patient in the hospital a short time ago, I overheard a nurse asking another nurse, " How do you mix Colymycin with saline for inhalation ? ... This stuff [sic] is starting to cake". The other nurse replied, " Ask one of the cystics.. they know how and they'll tell you ".

One patient, a 31 year old female { a college graduate who worked in a hospital setting } told me that she felt very uneasy about the whole idea of being treated on a hospital unit intended for adolescents. Specifically, she noted that one aspect she found " totally unacceptable " centered around the physical examination and questions she was asked by the resident and intern assigned to the unit. 15 { When patients were initially brought to the unit they were greeted by the floor resident accompanied by an intern. } Among

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15 Even patients admitted and treated by a private physician, were given a physical and asked a battery of questions by the resident and intern on the floor, and a nurse.

the questions asked of all patients are the following:

> " Do you drink alcohol ? Do you smoke cigarettes? Do you smoke marihuana ? Do you take drugs ? Are you sexually active ? Do you use birth control ? If so, what type ? What are your living arrangements ? How do you get along with other members of your family ? Do you have any friends ? Do you have any hobbies ?

This woman felt quite strongly that some of these questions ought not be asked of adult patients on the unit. Her objections not only centered on some of the questions asked of patients , but on the physical examination itself. Residents and interns are given an admission form { four pages long } which lists questions to be asked of patients, and it also outlines the physical examination to be performed. A resident physician told me, " Technically, we are supposed to examine the patient from head toe, but there does exists some variation in terms of how thorough we are ". This young woman { who has many hospitalizations for CF } told me that some of the physicians perform a real quick physical, while others are very thorough. This woman, Candy { psuedonym } told me that she felt bothered by the fact that the persons examining her were pediatricians and not

internists. 16

Another CF patient explained that while the doctors on the floor know much about cystic fibrosis, they know very little about other things relevant to older patients. He told me that he had renal disease arising from the cystic fibrosis, and that he was followed by a nephrologist. He said that he asked one of the residents a few questions about the medication { Capoten } he was taking for his blood pressure and found that the resident physician was unfamiliar with the drug. In the words of this patient, " The doctor told me that he was not a nephrologist and that the problems he was having with his kidney are seldom seen in kids { sic.} ". The patient stated that later, he asked his uncle, who was a physician, questions about the medication. When he told his uncle about his encounter with a physician who knew little about the medication, his uncle, replied " That drug is so widely used for the treatment of high blood pressure and renal impairment, that virtually any physician

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16 Since this is an adolescent unit, the house staff consists of pediatricians and residents specializing in adolescent medicine.

treating adult patients has experience with it ". This example represents one important limitation of treating older patients on hospital units designed for younger patients.

A 22 year old male CF patient who was a "regular" stated that he felt some of the nurses were very opinionated and overly judgmental. He felt that older patients on the unit were not accorded the respect that they should be given. He also stated that this perspective was not confined to the nursing staff but was to be found among physicians as well. He offered the following example to support his position:

> " The other night, my IV blew. Sharon {resident} came in and tried to start an IV. She stuck me three times and still couldn't get a vein. --- Twice she missed, and the last IV infiltrated a few seconds later. She told the nurse to page Ken { another resident }. About 20 minutes later, Ken arrived. He tried twice to get an IV going, and with no success. He proceeded to try a third time, when after having been stuck five times by now, I told him " forget it... no way ! ". He raised his voice and said, " You have no choice in the matter... just give me your arm. It's 11:30 PM ... I have better things to do than this.... just give me your arm !" I rang the buzzer and yelled for the nurse. Gail { nurse } came in. When I told her the situation she told Ken, " You can't force him to have an IV.... He's an adult. Ken left the room. A short time later a pediatric anesthesiologist { who was paged }

> started my IV, and he got it on the first shot. When I discussed the matter a short time later with Gail, she said, " What do you expect... this is a pediatric floor... he doesn't deal with many older patients ".

This adult patient believed that this whole encounter would not have taken place if he were placed on an adult unit, and if the physicians were more skilled at venipunctures.

Not all of the patients I encountered had reservations or negative sentiments about being treated on an adolescent unit. One man, recently married and with children of his own, told me that he " loved " being treated on the adolescent unit as opposed to being assigned to a floor in the main building. He stated that he found the time went by quickly since he was constantly involved in playing video games and socializing with teenagers and adolescents, as well as other CF patients. He remarked that the social worker who deals with CF patients told him " You are a role model and an inspiration to younger CF patients.... younger people who have CF see you as representing hope.... it reaffirms in their minds the notion that they too can live into adulthood with their

disease ". 17

One 17 year old CF patient told me that he was really pleased being on this unit because the nurses are not only more friendly, but they're attractive as well. A number of nurses on the unit are in their early or mid 20's. { Some of the nurses are single or recently married.} He somehow viewed this as an added benefit.

Stresses for CF Patients:

In spite of overall satisfactory relations between patients and staff, and the various parts which made one's stay more acceptable, stress arises. Many of these stresses are due to the unpleasant aspects of the medical treatment prescribed for the patient. In addition, hospitalized patients soon become accustomed to the routines of hospital life.

Most of the CF patients were on " daily weights." This means that every morning at 5:30 A.M. they were

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17 These remarks are paraphrased based upon the discussion I had with this patient. The reader should note that this is the same individual quoted in the anectdote on pg.97

awakened by the nurses aide who asked that they go to the bathroom to empty their bladder, and then step onto the scale. Shortly thereafter, they are greeted by the nurse who hands them their inhalations and then proceeds to perform drainage. Some people like to sleep and it is conceivable that this practice could disturb some people.

Between 6:30 and 8 A.M., the physicians caring for CF patients usually make their daily rounds. The thrombologists also make their rounds at this time, and bloodwork that has been ordered and signed for at the desk is dawn from patients. Shortly thereafter, the interns and resident physician on the floor make their rounds. It is not until about 10 AM that patients can get any rest without being bothered by the physicians. During the remainder of the day much time is spend between inhalations, postural drainage, and the administration of antibiotics. Many CF patients are on " I & O". " I & O " stands for " input and output. This means that all of the fluid intake and output of the patient needs to be charted. This information is of vital importance in terms of monitoring renal function.

As noted previously, many of the antibiotics that are used in the treatment of cystic fibrosis have serious side effects. As a result, " I & O " as well as frequent testing of blood { SMA6, CBC, Retic, platelets, BUN, Cretainine, } is common practice. In addition, patients often undergo frequent venipunctures to secure blood samples to determine " peak " and "trough" levels of aminoglycoside antibiotics they may be receiving. This means that the patient is stuck once to get a "baseline" blood value, the patient then receives the Tobra or Gentamicin within a one hour period, shortly thereafter a second blood sample is drawn, and about 1-2 hours later, a third blood sample is drawn. Cystic fibrosis patients like most other patients are not especially pleased by the prospect of having to be stuck with a needle three times within a three hour period.

Another problem that causes major upset for some CF patients is poor venous access. Some patients have " very good veins" ; it is very easy to start an I.V. on the patient, and the I.V.'s that are put in place usually run well, do not become infected, and last a few days. Unfortunately, not everyone is blessed with

" good veins." When veins " infiltrate " or " blow " , this is often stressful and traumatic for the patient. The greater the dosage and the more frequently the medication is administered, the greater the likelihood that the veins will blow. There is also a strong tendency for all of the veins on a given arm to blow simultaneously. When this happens. it is often difficult for an intern or nurse to establish another I.V. Also, often when a vein blows it becomes red and swollen and sometimes, phlebitic. Since CF patients are treated every 6-8 hours with three different antibiotics, and in doses which generally exceed the " usual adult dosage " { as is necessary to treat respiratory infections in CF patients } it is a common sight to see CF patients in torment because most of their veins are blown.

Many patients with cystic fibrosis also suffer from complications or other illnesses which are secondary to cystic fibrosis. For such patients, hospitalization is not a particularly enjoyable experience. For example, the patient who has CF complicated by diabetes must look after both con-

ditions. In the hospital setting, such patients are frequently subjected to " fingersticks " to measure their glucose levels, and they may have to follow a specific diet. Likewise, the care of the CF patient who suffers from liver disease or renal impairment often presents problems. Such patients must be carefully monitored, and frequent adjustments to their medications may be necessary.

Summary:

In this section I have examined the situation of the CF patient who requires hospitalization. Although CF patients may require periodic hospitalizations, the actual time spent by CF patients in the hospital varies considerably. Some patients seldom or rarely require hospitalization, while other patients require hospitalization more frequently. Based on conversations with some CF patients, it would appear that young adults with CF spent an average of two or three relatively short stays in the hospital. Since adult patients with cystic fibrosis are treated on an adolescent unit, I have inquired into patients feelings about this practice. I found that some patients had reservations

about this policy, while other patients found it not only acceptable but desirable.

I have also examined and focused attention on the specifics of what I.V. antibiotic therapy holds in store for CF patients, and what inpatient care is like for cystic fibrosis patients. By focusing specifically on the everyday functioning of the adolescent unit of the hospital, I have attempted to provide the reader with some sense of what hospitalization entails.

Chapter 6: Death in Cystic Fibrosis

> Death may be the greatest of all human blessings.
> ....Socrates in Plato's Apology

Awareness of Death in CF

Some CF patients are aware that imminent death is a distinct possibility. For a number, death is a real threat --- the likelihood of its occurrence is forever present. Most CF patients whom I encountered did not appear to be preoccupied with the subject. However, on occasion I came in contact with some young adult CF patients who spoke openly about death and dying. Such discussions tended to originate from one of the following: (1) during the course of hospitalization, (2) a downward shift in the patient's own health, or (3) from " outside forces."

Awareness that death could strike at anytime, and anywhere { including the hospital } frequently surfaced while patients were hospitalized. As Stadnyk [1973] states:

> "During hospitalization they (CF patients) see more severely involved patients. Such a patient, perhaps close to their own age, may die during their stay: and much of the patient's hope is

then destroyed. " 1

Stadnyk [1973] continues:

> "As they beome less able to function independently due to the decrease exercise tolerance they are further pushed toward dealing with the fact of their impending death." 2

Discussion of death and dying occurred in hospital settings under completely different sets of circumstances. For example, some young adults were involved in computer video games, when one patient remarked that a former patient was a "pro" at a game called " Dragon's Castle." The patient, noted in passing that that patient had CF and had passed on. For a few moments, discussion of how the patient died (ie, the nature of CF) ensued in a solemn manner. Minutes later the subject was quickly changed and involvement in the computer game continued as usual.

A second source that contributed to a patient's awareness of death was the onset of complications or a major shift in a patient's own condition. Patients who

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1 Stadnyk, S. " The Team Approach to Death and Dying", in Patterson, Denning, and Kutscher, op cit, pp. 134.

2 Ibid, pp. 134 ff.

suffered marked hypoxia or who had a pneumothorax could conceivably wonder about the possibility of their own death. As a patient becomes less able to carry out minor tasks (such as walking, eating, etc.), it is likely that he/she might wonder if they are passing through the stages that lead to one's demise.

Another source that results in a patient's awareness of dying are " outside forces." For example, the National Cystic Fibrosis Research Foundation aired on public television commercials designed to generate funds for CF research. One commercial featured a young child with cystic fibrosis who was " hooked up " to monitors with an IV running, while he was receiving oxygen via a mask. The child appeared to be lifeless. The portrayal was accompanied by a narrators remarks and pleas which stated, " If you have CF, chances are you won't live past 20 " (paraphrased). This commercial disturbed and offended many CF patients and their families. First, the narrator's words were not entirely accurate, and secondly, it served to reinforce in some CF patient's own minds their awareness that cystic fibrosis is a serious, deadly disease. While advertising and solicitation may be necessary to secure

funds for research, responsibility and proper judgment are also prerequisites for accomplishing this goal. Many CF patients and members of their families felt that this particular commercial and similar tactics went too far. In the minds of some CF patients the commercial exploited the unfortunate situation of CF victims. A New York State couple with two young CF children brought the Cystic Fibrosis Foundation to Court in an attempt to terminate the airing of the commercial. The couple unfortunately lost their case. The couple had emphasized and instilled in their children's minds the idea that they could live a normal productive life in spite of their physical handicap. The mother of the children argued that this commercial certainly seemed to call that philosophy into question. The result of the Court's ruling was fourfold: (1) many Cf patients were unhappy or offended by the commercial. As one CF patient recalled, " At the time, I felt a sense of numbness whenever the commercial was aired." (2) The CF Foundation did not raise the level of funding it had hoped to { although it should be noted that the Cf Foundation claimed that it resorted to such tactics because of a decline in funds over

previous years), (3) some of the strongest financial backers of the CF Foundation are CF patients, their families and friends. In addition to contributing their own money, family and friends of CF patients are largely responsible for seeking contributions from the general public. Since some of the patients and their families felt deeply hurt and alienated by the actions of the CF Foundation, some people did not feel overly enthusiastic towards the CF Foundation, and (4) perhaps most damaging, the commercial heightened and magnified in CF patient's minds the consciousness that there is no cure for CF, and that death from the disease is unavoidable. This endeavor perpetuated by the National Cystic Fibrosis Research Foundation clearly had many drawbacks in the eyes of some CF patients.

Patterns of Dying:

Glaser and Strauss [1968] discuss the process of dying employing what they term " trajectories " of dying. Some deaths occur instantaneously, while others, which Glaser and Strauss refer to as " lingering trajectories ", progress at a much slower rate. According to Glaser and Strauss , the dying trajectory of each patient has at least two properties --- duration and

shape. Duration refers to the time period leading to the patient's death, while " shape " refers to the course of the patient's death.3 Some patients deteriorate rapidly and are subjected to a straight downhill progression of their disease prior to death. Other patients facing death may lose ground, improve slightly, and then gradually lose ground prior to their death. The " shape " of a trajectory of dying can, therefore, be graphically projected. In cystic fibrosis, the progression of the disease varies from one patient to the next. Some patients decline rapidly and die during the first few days of life or in early childhood. Others survive into the teens and beyond.

Medical authorities queried suggested that death in cystic fibrosis tends to follow a "typical" pattern. Belmonte and St. Germain [1973] present the following description of the dying process as it occurs in cystic fibrosis:

> "Death in cystic fibrosis is slow in coming and extremely painful to watch. The child has be-

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3 Glaser,Barney and Anselm Strauss, Time for Dying, { Hawthorne,New York: Aldine Pub. Co, 1968, p. 150 ff.}

> come progressively weaker and more short of breath. The pulmonary embarassment increases as he/she is less able to cough and bring up sputum. CO2 retention and anoxia lead to somnolence mixed with anxiety and agitation. Real fear develops as respirations become progressively more labored and anoxia more severe. At this point and preferably at its onset one should resort to the judicious use of sedation to ally fear by sleep. Still the struggle to breathe persists in spite of the stupor and there may be 10-24 hours more ahead before death finally supervenes...." 4

This description suggests that the trajectory of dying associated with cystic fibrosis is usually one of lingering. It is slow in its arrival and gradually progresses until death ensues.

While medical authorities are not hesitant to describe the process of dying in cystic fibrosis, it is important to realize that their descriptions and information are limited and only somewhat accurate. They only describe the events of the patients final days before death. In cystic fibrosis, as is the case in many illnesses, there is considerable variability in terms of the period of time from when a patient begins " losing ground" and fails to get better, until he/she

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4 Belmonte, Mimi and St. Germain, Yolande, " Psychosocial Aspects of the Cystic Fibrosis Family " in Patterson, Denning, and Kutscher, op cit., pp. 88.

is entering the final phase of the disease. Some patients may suddenly, in a matter of days or weeks, suddenly become very ill and die. Other patients, may be quite ill for a long period of time, and after a year or longer of not responding to medical care, they may experience a medical crisis such as a pneumothorax, a serious respiratory infection, or they may develop total resistance to all available antimicrobial agents, and then death may follow.

I observed one case in which a 16 year old CF pat- had spent close to a year in the hospital on a continuous basis { One nurse told me that he only spent a total of about ten days over the past year, at home }. The patient had developed repeated bouts of Pseudomonas pneumonia, and the organisms he possessed were virtually all resistant to every antibiotic available on the market. { According to his mother, even the quinolones, a new group of antibiotics which at the time the FDA released for " investigational use only ", failed to provide any benefit }. In summary, this represents an example in which it took death a substantial period of time to catch up with the patient.

Demands on the Family:

Poss [1981] maintains that the activities placed on a family facing the loss of a loved one, can be viewed as " a set of linked problem - solving tasks, at least some of which need to be resolved if the crisis is to be mastered ". 5 After reviewing the literature regarding demands placed on the family facing an impending death of a loved one, Poss found the following " tasks " tend to be elaborated upon frequently:

1. responding to whichever terminal crisis task their dying relative is engaged in, that is, as corollaries or complementary actors in the situation.
2. reviewing their lives together
3. evaluating their relationships with the dying person and, as a follow-on, reflecting on themselves and on their own lives.
4. dealing with unresolved business in the past.
5. completing any unfinished issues.
6. caring for patient in the present.
7. participating in decision regarding life-prolonging procedures for the patient.

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5 Poss, Sylvia, Towards Death with Dignity: Caring for Dying People. { National Institute Social Services Library, London, George Allen & Unwin Ltd, 1981, p.39 }

8. using their now shortened time together as creatively as possible in the present.

9. balancing their energies, and resources between their own adjustment and their care of the patient and between issues of the past, present and future.

10. handling their guilts in relation to the patient's dying and then his death.

11. planning for the future without the patient.

12. making new resolutions about their own lives as a result of their contact with loss and death.

13. reassessing relationships with God in the light of their encounter with death.

14 realigning the disrupted family system after death.

15. grief work, including experiencing the intense pain of bereavement, emanacapating themselves from bondage to the deceased person, readjusting to the environment in which the deceased relative is missing and forming new relationships and patterns of rewarding interaction. 6

Although many of these elements do not apply to younger CF patients, they may be relevant to some older CF patients.

Belmonte and St. Germain note that the observation

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6 Ibid, p. 39

of a CF patient dying is extremely painful to watch, and may be intolerable for parents of a dying patient:

> ".... The plight of the parents at this time is almost unbearable. Every effort should be made to insure that the child is not conscious and that the parents are aware that this is so.... The last hours of the dying CF child may be intolerable for some parents to watch. The social worker and physician should be alert to these feelings and be prepared to give support should the parents prefer to stay away. In this case a substitute person should be found to remain with the dying child." 7

The physical, emotional, and psychological demands placed on the family of a dying patient can be a heavy burden to bear. Since the trajectory of dying in cystic fibrosis is often lingering, this can place further hardships and stresses on family members.

The Organization of Death and Dying:

While serving as a hospital volunteer a number of summers ago,I recall the death of a nineteen year old male who succumbed to cystic fibrosis. For a period of almost three days prior to his death, his parents and nurses who tended to him described his plight as involving intense suffering. The patient's father would

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7 op.cit, p. 88

spend a couple of minutes in his son's room, and then he would return to the smokers lounge located outside of the unit. There he and his wife spent endless hours. The man often wept while his wife appeared teary eyed and emotionally distraught. Both the husband and the wife looked as if they had literally been through Hell. Fellow family members and friends tried to comfort them but seemed to provide little consolation. The hospital rabbi was also summoned and he tried to do all that he could. What was more traumatic for the parents was that this was not the first time that they had lost a child. Two years earlier, at about the same time (July) they lost their other son who also died of CF. I remember at one point the man crying profusely and stating, " It's happening again... it's the same thing all over again, and I can't take it anymore " [paraphrased]. The man's weeping was so intense, that it was readily heard from the lounge area into the hallway leading to the patients rooms. Nurses from the unit quickly escorted the man and his wife to a staff lounge located at the opposite end of the hall. There was hardly a person on the floor who was not aware of the fact that a 19 year

old boy named Jonathan was dying of cystic fibrosis in the private room at the end of the hall.

In his work, Passing On: The Social Organization of Dying, Sudnow observed that Emergency Room staff routinely passed judgment on the " worthiness" of saving and tending to the dying patient.

Demands on Medical Staff:

Sudnow [1967} observed that since the 1960's more people die in hospital settings than ever before. As a result, nurses and physicians are likely to come in contact with patients who are critically ill and patients who are likely to die while under their care.

Many writers have noted that doctors and nurses experience feelings of frustration, anger, threat, guilt, impotence, despair, failure and fear in relation to their work with the dying patient. 8 It was my observation that interns and residents on the adolescent unit tried as much as possible to isolate themselves and avoid contact with patients who were dying.

A 30 year friend of mine who is a pediatrician

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8 Poss, op cit, p. 113.

told me that he always finds the loss of a child an emotionally grabbing event. According to him, part of it is due to the fact that physicians view pediatric patients as just starting out in life and as totally innocent victims and totally undeserving of the horrors that can happen in life. He also stated that many pediatricians tend to be young males or females who are in the process of starting a family, or they already have children of their own. He told me that when a child dies it is frequently upsetting to him because it reaffirms in his own mind the sanctity of life and the realization that death can strike his own 3 year old son.

The Hour of Death:

Two medical social workers have noted that for some unknown reason, it seems that most of the deaths occur during the night. 9 Many nurses that I spoke with seemed to agree. One nurse, about 25 years of age and a recent nursing school graduate, pointed out that

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9 Childress, Josephine and Harrison, Gunyon, " Working With the Parents of the Dying Child: A Shared Responsibility" in Patterson, Denning, and Kutscher, op cit.,p. 176 ff.

" night deaths" have some consequences. During the night and early morning hours there is less staff to attend to patients. There are usually two or three nurses scheduled to work the 8 p.m.- 8 a.m. shift. 10 In addition, generally few doctors are present on a regular basis on the unit. This means that should an emergency occur, nurses must beep (page) the doctor(s) on call. Nurses attend to the dying patient while the physician is summoned. Two nurses (whom I have known for many years, since I was a volunteer) stated that some interns and residents are especially slow in responding to pages during the nightime and early morning hours. One of the nurses suggested that this was purposely done, and she mentioned that frequently by the time a physician finally arrives in the patient's room, the patient is usually gone or totally uncon-cious. This was usually true whether the patient was dying of CF or some other cause. The same nurse remarked that after a patient dies, interns and

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10 The adolescent unit and PIC have recently switched to 12 hour shifts. As a result, the evening shift begins at 8 PM and ends after report at 8 AM, the next morning. Under this system, nurses work three days a week, and once a month they work an extra day to fulfil a 40 hour work week.

residents working the early A.M. hours typically emerged from the unit as soon as possible. Sometimes, physicians left immediately after nofifying a patient's family by telephone. Perhaps physician's fears of blame or responsibility from a deceased patient's family contribute to this practice. It is also possible that a physician's own sense of insecurity surrounding death, or " detached concern " [Fox] may be factors that are operative in these situations.

Summary:

In this chapter we have focused attention on the process of death and dying as it applies to the CF patient.

Cystic fibrosis patients suffer a slow and lingering death that is marked by severe hypoxia which results in difficulty breathing. As the patient's breathing becomes more labored, real fear and anxiety sets in. At this point, physicians often prescribe sedatives to make the person's impending death as comfortable and as peaceful as possible.

Based upon the available literature, as well as my observations in the hospital setting, it is apparent

that the predicament of a child dying of CF may prove too emotionally trying for many parents to handle. At such time, social workers suggest that a "substitute" person be found to stay with the child.

Nurses whom I spoke with indicated that many CF patients die during the very early morning hours { after midnight but, before 8 AM }. This is a time when few physicians are on the floor. Nurses I queried suggested that most interns and residents " on call " during these hours, are slow in responding to pages in which a CF patient is experiencing death. Consequently, many of the patients experience their deaths while being attended to by nurses as opposed to physicians.

Sometimes a peculiar phenomenon occurs regarding the reaction of family members to the death of a CF patient. I personally have witnessed situations in which parents and spouses of seriously ill CF patients have expressed their hopes that death would occur soon so that their loved ones would not suffer. Yet, in a number of cases, when parents were first told of the patients demise, they respond with shock, disbelief, and sometimes, a feeling of anger directed at the

staff. Statements such as " Doc, you couldn't do anything.... couldn't you try something else ! " or " I knew she/he was ill but I never thought I would lose her... not so soon " were sometimes directed at physicians. It is interesting to note that patients and their families are not hesitant to acknowledge the seriousness of the disease, yet, when a patient dies they seem as though the patients death was a total shock. It seems as if persons close to the dying patient know that death is imminent, yet they have difficulty accepting its occurence. Perhaps, in this regard, Waddell is correct when he claims that the process of "neutralisation" is operative in cystic fibrosis.

Each patient with cystic fibrosis reacts differently to the prospect that they will succomb to their disease. While some patients may shy away from the reality that some day they too will die of CF; others are constantly reminded that life is precious and it is something that can never be taken for granted. As one 28 year old doctor who has cystic fibrosis and who renders care to CF patients was recently quoted as saying, " It's very hard, when you see what others go

through in the final stages, not to project what might be in store for you,".... " It is frightening, and I do my best not to think about it." 11

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11 Abrams, Arnold, "The Doctor Has the Disease Too ", NEWSDAY, February 19, 1986, Part II, p.3

Chapter 7: SUMMARY AND OBSERVATIONS

Reflections:

In the course of my fieldwork, a number of crucial themes in the sociology of medicine often arose. These included: the quality of life for CF patients, the quandary of catastrophic illness, pain and suffering endured by CF patients, faith and hope, CF patients reliance on experimental drugs and advances in medicine, tragedy and despair, and, death and dying.

Two of these themes, tragedy and despair, and, death and dying, were especially emotionally moving. One situation which left its mark in the minds of many people was the following:

> A nine year old boy (who had CF} and his eleven year old sister came home from school and found a note addressed to their grandfather. By nighttime, when their father had not returned home, the girl telephoned their grandfather. Two months earlier, the children had lost their mother to Cancer.
>
> The father left a note in which he stated that the loss of his wife was a traumatic experience, but the thought of losing his son from CF was more than he could emotionally handle at the moment. He asked that the grandfather { his father } look after the children until he could sort things out.
>
> Under the circumstances, the grandfather felt he had no choice. { He had recently

> retired due to poor health. He returned to work, as a salesman in a furniture store }. Although there was no telephone contact from the father, he regularly sent letters to his family and certified bank checks of considerable amounts of money. He asked that the monies be used to provide for his children, and especially for his son's medical treatment.
>
> Following the death of his mother, the boy's pulmonary condition deteriorated rapidly. A short time later, he developed a hernia. While being operated on for a hernia, the boy went into respiratory arrest and died on the operating table. At the age of nine, CF had claimed another victim.

A number of CF patients also experienced similar tragedies in their own lives. Some patients had siblings who were born with CF, but who had died of the disease. One 26 year old woman {now deceased} lost her mother to Cancer a short time ago. In addition, her father was an alcoholic. Two other CF patients lost their fathers to heart disease when their parents were in the early 50's.

In addition, a number of the young adults with CF were critically ill, and either during my research or shortly thereafter, they expired from CF. This served to constantly reinforce in my mind the fact that CF is a degerative disease which inevitably leads to death.

Alternative Approaches:

Medical sociologists are interested in quasi-healers, folk medicine, and alternatives to orthodox medical treatment. Alternative approaches to medicine are important because they allow patients who are disheartened by traditional medical care a possible alternative. Since treatment for CF is primarily palliative care rather than curative, alternative modes of treatment may provide appeal to some patients.

Within recent years, some patients have decided to sway from traditional medical treatment for CF, and they have turned to other forms of treatment. Joel Wallach, a one time researcher for the Cystic Fibrosis Foundation, { and a veterinarian } represents one such alternative. Wallach claims that cystic fibrosis is caused by a deficiency of the mineral selenium. He has a sizeable following of CF patients who follow a rigid dietary regimen which is high in selenium and other vitamins, and provides for a high caloric intake. While many physicians in cystic fibrosis channels are critical of Wallach's " treatment ", some physicians believe that his approach is not totally

without merit. At the present time many of his followers reside in the Western region of the United States. As reported on National television { The ABC " 20/20 " News magazine show } a short time ago, a number of CF patients who have followed the Wallach protocol report substantial improvement in their pulmonary and gastrointestinal status. Wallach's approach is of key importance in that it allows those patients who are displeased with " orthodox " medical management of their disease, an alternative.

More CF Births:

At present the incidence of cystic fibrosis is growing. A decade ago, 1 in every 1,200 - 1,500 live births in the United States yielded a CF victim. At present, the incidence is approximately 1 in every 950 - 1,000 births. If this trend continues, it is likely to be advantageous for CF patients. As more persons are born with the disease, more research funds are likely to be generated. In addition, greater production of the medicines used primarily by CF patients is likely to yield a decline in their costs.

An increase in the number of CF patients will result in a greater demand for pediatric pulmonary

physicians and physicians with training in critical care medicine.

Self Help Groups:

At present most self help groups for CF patients operate on the local level { ie, they exist as small groups of older CF patients who meet on an irregular basis within the confines of the hospitals where they receive care }. If the life span of CF patients continues to rise. coupled with increased numbers of patients, it is likely that there will be an expansion in the networks of CF self help groups.

Need for More Government Funds:

If the plight of CF patients is to improve substantially, it will be necessary for far more money to be allocated for medical research. Moreover, the costs of existing care must fall within the reach of all CF patients. At the present time, there are CF patients who have relinquished treatment for their disease because they lack money to pay for their care. As a result, these unfortunate souls are suffering needlessly as well as diminishing their life span.

Many states have recently established State Programs for CF patients. Unfortunately, many of the programs such as the New York State CF Program are administered by heartless bureaucrats whose primary interests lie in saving money. { I know of one case in which a patient was shifted back and forth between Medicaid and the New York State CF Program. In the end, he had coverage from neither, and he was only able to acquire a basic Blue Cross hospital plan }. This patient had to eliminate receiving medicines, oxygen, and other care for CF. This case does not represent an anomaly. In the course of conducting interviews, I came in contact with older patients who told of exhausting their resources and of having to sacrifice their care. If this trend is to decline, more government financial support of substance will have to be allocated.

Need for More Behavioral-Social Science Research:

The bulk of the existing literature on CF concerns the disease as it relates to children. This is due to the fact that until recently, few CF patients survived past the early childhood years.

As discussed in Chapter 1, most written mater-

ials regarding cystic fibrosis concern biomedical and medical aspects of the disease. There are a few articles written about behavioral and familial coping patterns in cystic fibrosis. Nevertheless, the existing literature is seriously lacking. To date, the most popular book on psychosocial aspects of CF is Psychosocial Aspects of Cystic Fibrosis: a model for Chronic Lung Disease, edited by Patterson, Denning, and Kutscher. The book was published in 1973 ---- 16 years ago.

There is a compelling need for this literature to be updated and expanded and to pay closer attention to the needs of older patients with cystic fibrosis.

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