Family Adjustment to Living with Cystic Fibrosis

by Gretchen Lanka Allen

Cystic Fibrosis (CF) is an intriguing disease that raises myriad issues relating to health, wellness, treatment and adjustment for families living with the disease, as well as for healthcare and human-services professionals working with these families. To begin to understand this complex illness, we must explore the biological, physical and psychosocial needs and strengths of families living with cystic fibrosis. In addition, we need to look at how living with CF can affect child and family development and the adjustment into adolescence and beyond. This paper begins with an introduction to the Family-Systems-Illness Model, an adjustment model that provides a clear framework for social workers and other health care professionals working with clients with CF. The paper then looks at cystic fibrosis, its etiology, symptoms, course and prognosis. It then discusses how the particular features of CF can affect family and child/adolescent development. The paper continues with a discussion of the Family-Systems-Illness Model, and it ends with some questions that are important for a social worker to include when assessing a family caring for a child with cystic fibrosis.

INTRODUCTION

Cystic fibrosis (CF) is a complex and intriguing disease that raises myriad issues relating to health, wellness, treatment, and adjustment for families living with CF. While health care professionals and researchers continue to make significant steps to improve and extend the lives of patients with CF through experimental gene therapy, new medication treatments and improvement in physical therapy, there is still no cure (Cystic Fibrosis Foundation (CFF), 2002). Children, adolescents, and young adults with CF can live healthy, active and productive lives. However, without a cure, those living with CF always will have it and most will have a shortened life span. Until a cure is found, the best that doctors can do is to relieve the symptoms of CF so that individuals can live with an improved quality of life (National Heart, Lung &

Blood Institute (NHLBI), 2002).

To begin to understand this complex disease, we should have a framework or model through which we can assess and understand this disease and those affected by it in a holistic sense. We must explore the biological, physical and psychosocial needs and strengths of those living with cystic fibrosis and their families. In addition, we need to look at the ways in which living with CF can affect child and family development and the adjustment into adolescence and beyond. I begin this paper by introducing the Family-Systems-Illness Model, a normative model that provides a clear framework for social workers and other health care professionals working with clients with CF. Next, I describe cystic fibrosis, its etiology, symptoms, course and prognosis. I then discuss the particular features of how CF affects family and child adjustment through child and adolescent development. I end the paper with a discussion of the adjustment model that provides a clear framework for health care professionals working with patients with CF, along with some questions important to include when assessing a family caring for such a child.

INTRODUCTION TO THE FAMILY-SYSTEMS-ILLNESS MODEL

The Family-Systems-Illness Model was developed by Dr. John Rolland through his work at family health centers in New Haven, Conn., and Chicago. It is a normative model that describes the complex interactions between a particular physical illness or disorder, the patient or client and the family as a whole (Rolland, 1994). Often families referred for psychological or psychosocial help during an acute physical illness have never seen a mental-health professional. Some sources cite that up to two-thirds of families facing serious illness fit into that category, underscoring the importance of having a comprehensive model we can use to assist the average family coping with some common and expected stressors of illness (Rolland, 1994). The Family-Systems-Illness Model provides just that: a multi-systemic view of the many interactions and interactive processes between the demands of different illnesses over time and key parts of family functioning.

The model looks at family style, cohesion and adaptability through a "strengths" perspective, examining both a family's strengths and its vulnerabilities. It offers a useful, systemic view of family adaptation to serious illness in a child as a normal and expected developmental process over time (Rolland, 1994). The model also provides social workers and health professionals with a conceptual base for approaching practice from a family systems perspective,

emphasizing the initial consultation and family assessment as well as the common issues and challenges that families face as they experience a condition over time (Rolland, 1994, 12). The model is useful when trying to understand and treat patients with severe illness and disability because it is interdisciplinary, and it can be used and applied by both health and mental-health workers, from physicians to social workers. The model is particularly well-suited to social workers within the health and mental-health environments because it takes into account various spheres of influence that affect family adjustment and development. The model moves within the broader sphere of belief systems, culture and ethnicity, to individual, family and illness life-cycles, and into the specific and precise type of illness, adversity, or loss (Rolland, 1994, 14). With this multi-systemic framework in mind, we can begin to look at the complexities of cystic fibrosis and begin to understand why the Family-Systems-Illness Model can help us understand families coping with this disease.

DESCRIPTION OF CYSTIC FIBROSIS

Cystic fibrosis is a chronic, progressive and frequently fatal genetic disease affecting some 30,000 Americans and their families (CFF, 2002; Rolland, 1994). Each year, around 2,500 babies are born with CF in the United States. Most babies with cystic fibrosis are born to families of Caucasian, northern European heritage, although CF affects all races and ethnic groups. In addition, around one in every 31 Americans (one in 28 Caucasians) is a healthy, unaffected carrier of an abnormal "CF gene." In other words, some 12 million Americans unknowingly carry the gene that causes cystic fibrosis. This prevalence makes CF the most common hereditary disorder in America (CFF, 2002; Hollander, 1997; NHLBI, 2002). The average person with cystic fibrosis lives into early adulthood. According to recent data, the current median life span for people with CF is 33 years (Desrosiers et al., 1996).

Cystic fibrosis mainly affects the respiratory and digestive systems in children and young adults. A child who inherits two CF genes from his or her parents has a defect in a protein that regulates the levels of sodium and chloride (salt) within cells lining organs such as the lungs and pancreas. A defect in this protein causes a person with CF's exocrine glands to absorb too much water and to produce abnormally thick and sticky mucus that can clog the lungs, pancreas, and other organs. This thick mucus can cause frequent lung infections and often prevents pancreatic enzymes from reaching the intestines to help break down and digest food (CFF, 2002; Silverstein et al, 1994).

CF can lead to various other medical problems, as well. The most common medical problems for people with cystic fibrosis include: sinusitis; nasal polyps (growths inside the nose); "clubbing" or rounding of the fingers and toes; pneumothorax (a tear in the lung tissue causing trapped air in the chest wall); coughing of blood; *cor pulmonale* (enlargement of the right side of the heart); abdominal pain or discomfort; rectal prolapse; and sterility in 98 percent of men. Some people with cystic fibrosis may also develop liver disease, diabetes, pancreatic inflammation or gallstones (NHLBI, 2002).

Cystic fibrosis has a variety of symptoms and does not follow the same pattern in all patients. While the majority of children with CF are diagnosed before the age of 2, some patients reach adolescence or early adulthood before receiving a correct diagnosis (Hollander, 1997). CF symptoms can seem similar to those of asthma, chronic bronchitis, or gastrointestinal disorders like celiac disease. The symptoms of CF affect different people to varying degrees in different ways (NHLBI, 2002). The most common symptoms of CF are salty-tasting skin, a persistent coughing or wheezing, excessive appetite with poor weight gain, or a "failure to thrive," and bulky, foul-smelling stools (CFF, 2002). Occasionally, babies with CF are born with an intestinal blockage called meconium ileus. In those cases, the infant can be diagnosed at birth.

The test most often used to diagnose cystic fibrosis is the sweat test, a simple and painless test measuring the amount of salt in the sweat. The sweat test may not work well with newborns who do not produce enough sweat, and a small percentage of children with CF have normal amounts of salt in their sweat (CFF, 2002). Patients also can be diagnosed using chemical tests for the presence of the mutated CF gene, or through chest X rays, phlegm cultures, stool examination and lung function tests (NHLBI, 2002). After a positive diagnosis, the treatment for CF depends on the stage of the disease and which organs are affected. Cystic fibrosis is most often treated with antibiotics, to cure lung infections, and a combination of physical therapy, exercise and medications to treat and clear the accumulated mucus that coats and blocks the airways (Pulmonology Channel, 2002). The most commonly used treatments are airway clearance techniques (ACT) such as chest physical therapy (CPT), in which a caregiver or therapist uses vigorous percussion with cupped hands on the back, sides and chest to dislodge mucus from the lungs (CFF, 2002). This treatment is also known as chest percussion, thumping, clapping, bronchial or postdural drainage or chest physiotherapy.

Patients with CF also can use various breathing techniques, a hand-held flutter device that causes vibration to remove mucus, or an inflatable therapy vest, also known as high frequency chest compression (Pulmonology Channel,

2002). The breathing techniques, flutter machine and inflatable therapy vest all allow those living with CF to have greater autonomy and self-reliance. Adolescents away at school, or young adults in college or living on their own, often will use these techniques and devices for self-care (Desrosiers et al, 1996; Hollander, 1997; Lubich, 2002). In addition to chest physical therapy, it is important for children and adolescents with CF to exercise regularly to increase and maintain lung functioning. They also need dietary and vitamin supplements and often have to take pancreatic enzyme tablets before eating to aid digestion.

The course of cystic fibrosis is unpredictable. Most children with CF will need to have medical checkups every three months to ensure continued lung function and general health. Some children remain relatively symptom-free while others need frequent medical care and "tune up" visits to the hospital to stay alive (Harris & Super, 1995; Shapiro & Heussner, 1991). During the life course of a child or adolescent with cystic fibrosis, she will have frequent lung infections, changes in lung function and problems with digestion. At times her daily treatments are sufficient to maintain her health; at other times she will need adjunctive therapy. A child with cystic fibrosis may need extended hospital visits or may get by on occasional checkups. In addition, some children and adolescents with CF take up to 20 medications a day, while others rely mostly on physical therapy and exercise (Hollander, 1997; Lubich, 2002). The variability and unpredictability of the course of cystic fibrosis can mean different things for each individual patient's prognosis.

Technological advances over the past 50 years have raised the median life span for a person with CF from less than eight months in the 1940s, to 11 years in the 1960s, to the current average life span of 33 years (Desrosiers et al., 1996). This means, however, that some people with CF die in childhood while others may live into their 50s or 60s. Some patients who receive successful lung, pancreas, heart and liver transplants are enjoying health they never knew. While the course and prognosis for people with CF can be varied, cystic fibrosis is nonetheless a fatal disease. According to current literature, CF is described as a disease that leads to a "shortened life span" (Rolland, 1994). Most patients with cystic fibrosis eventually die from respiratory failure or *cor pulmonale*, an enlargement of the right side of the heart. About 95 percent of people with CF die from lung infections (Pulmonology Channel, 2002). That sense of inevitability, anticipation of grief and the highly unpredictable nature of the disease can lead to countless difficulties for children and families who have to continually adjust and readjust to living with cystic fibrosis.

FAMILY/CHILD ADJUSTMENT AND DEVELOPMENT

Although individuals and families living with cystic fibrosis can have active and healthy lives, there is no denying the amount of time and energy those affected have to spend on daily treatment, physical therapy and the many psychosocial issues associated with CF. Children and families living with cystic fibrosis face numerous additional daily stressors. Many features of living with cystic fibrosis can have a deep impact on the psychological functioning and adjustment of the child with CF, other family members and the family as a whole. In particular, CF's chronic nature, uncertainty about long-term prognosis, the genetic link and the great impact on daily family life all affect functioning and adjustment (Harris & Super, 1995; Rolland, 1994). Like other chronic diseases in children, cystic fibrosis can cause a host of problems for the family and individual living with this disease. The additional burden on the family posed by dealing with cystic fibrosis may lead to marital conflicts, limit social life, siphon attention away from healthy siblings and drain financial resources (Shapiro & Heussner, 1991).

Since most CF diagnoses are made in early infancy or childhood, the parents, caregivers, and siblings are usually the first to experience psychological distress (Harris & Super, 1995). After the initial shock and grief, the most marked change for a family with a new diagnosis of cystic fibrosis is the amount of time and energy necessary for "home care." Families caring for a child who has a new CF diagnosis will not have any preparatory time to adjust before they must begin the child's complex daily care routine. People with cystic fibrosis usually need three to five hours of daily medication and physical therapy to maintain health (Hollander, 1997; Desrosiers et al, 1996; Pulmonology Channel, 2002; Shapiro & Heussner, 1991). These particularly rigorous treatment demands can be exhausting, and the various treatments and medical devices necessary to maintain health and improve quality of life can be prohibitively expensive.

Most children diagnosed with cystic fibrosis adapt well to the diagnosis and need to spend little time in the hospital (CFF, 2002; Harris & Super, 1995). When they learn the home-care routines necessary each day, they are generally flexible and able to incorporate treatment as "just part of the day" (Hollander, 1997). However, certain developmental issues and difficulties often arise for those with cystic fibrosis and their parents or caregivers. The rigorous treatment demands and amount of time and energy most people with CF have to spend on maintaining health can be exhausting and overwhelming.

In addition, some aspects of the disease may be embarrassing for children

and adolescents. For parents of very young children with CF, issues like toilet training and "appropriate touching" can be problematic. Because children with CF often have large, bulky, and foul-smelling stools, toilet training can be awkward. As the young child with CF begins school and has to use public restrooms, the "stinky poop" that permeates the bathroom or hallway might embarrass him and subject him to ridicule (Shapiro & Heussner, 1991). Older siblings also may be embarrassed by the foul smell and may feel uncomfortable inviting friends over.

As communities become more open and aware about child abuse, it can seem horrible for parents to perform chest physical therapy on a small child. While the child learns in school that she shouldn't ever be hit and about "appropriate" and "inappropriate touching," she has to go through daily sessions of being hit on the back, chest and sides by adults. As a child becomes more aware and sensitive about his body, he can be uncomfortable with all of the doctors, nurses and physical therapists that examine, poke and prod him. In addition, a school-aged child with CF may be embarrassed about the number of pancreatic enzyme pills she has to take before eating a school lunch. Some children have been known to conceal or discard their capsules (Harris & Super, 1995). A child may also be resistant to having CPT sessions interfere with school or play time. Furthermore, a child who is sick and requires frequent hospitalization can fall far behind her classmates in school.

Because a child with cystic fibrosis can be particularly vulnerable to respiratory infections, parents may want to overprotect that child from being exposed to germs. This could mean keeping a child with CF away from other kids or keeping her inside so that she won't catch anything. Health providers must remind parents of children with CF that it is impossible to protect any child from all germs, and that a child's emotional and mental health and growth/maturity depend upon living "as normal a life as possible" (Silverstein et al., 1993).

As children progress toward adolescence, independence, physical appearance and behavior become paramount. Adolescents can become obsessed with their appearance, relationships with friends, and blossoming sexuality. This is a time of emerging independence, when peer groups and others' opinions become paramount, and parents become terribly "un-cool." These developmental issues can be particularly charged for adolescents living with CF.

In terms of behavior, while smoking or taking drugs can be devastating for anyone, they could seriously hurt or shorten the life of an adolescent with CF. While other teens experiment with smoking, a teen or child with CF cannot experiment in the same ways. This can lead to feelings of extreme isola-

tion and resentment of the disease, doctors, parents and others. In addition, a teen with CF may not be able to tolerate being around others who smoke, as the smoke could cause severe reactions such as coughing fits. Even if a teen with CF wants to fit in by "hanging out" with friends who experiment with cigarettes and drugs, he might not be able to. A teen with CF also may seek independence and autonomy with their treatment, and parents need to work with health care professionals to allow for some flexibility, if needed.

The most devastating "side effect" of adolescence on cystic fibrosis can be rebelliousness or resistance to treatment. Most teens with CF at some point try stopping their medications and daily therapy, but that rebelliousness can lead to disastrous and long-term effects (Lubich, 2002). While healthy teens may feel invincible and unstoppable, adolescents with CF have to face mortality at a young age. Having to be "old before your time" can take its toll on an adolescent's development (Hollander, 1997). At such times, teens with cystic fibrosis can benefit greatly from support from other teens with chronic illnesses. Most cystic fibrosis care centers will have some type of supplemental support system for teens in crisis.

While children and adolescents with cystic fibrosis may appear just like any other child, there are certain physical traits associated with CF (Hollander, 1997; Lubich, 2002; NHLBI, 2002). Many children with CF have trouble gaining weight and may be short and skinny. The "failure to thrive" they experienced around the time of diagnosis may continue and cause them to look smaller than their peers. In addition, a number of people with cystic fibrosis develop a barrel-shaped chest because air gets trapped in their lungs, causing the rib cage to expand. Some children with CF have puffy, "chipmunk" cheeks because of medication (Lubich, 2002). Additionally children and adolescents with CF may try not to cough because they are embarrassed about coughing all of the time, but coughing is necessary to loosen and bring up mucus to maintain healthy airways (Lubich, 2002; Silverstein et al., 1993, 41).

Dieting and food can be another developmental issue for adolescents with CF. Many people with CF need to eat a great deal of food to maintain their weight, sometimes ingesting upwards of 3,000 to 5,000 calories a day. While other adolescent girls may obsess about dieting and fat intake, a young teen with cystic fibrosis might eat a pint of ice cream and a 1-pound steak in one sitting. That discrepancy in dietary habits may cause girls with CF, in particular, to feel awkward or left out. One 14-year-old girl with CF commented that while she loves "getting to eat whatever I want, whenever I want," she sometimes gets embarrassed by the amount she has to eat in front of her friends (Hollander, 1997).

To help children, adolescents and families cope with these difficult aspects of adjustment and development, it is important for health care providers to have an understanding of the complexities of each family's adjustment. As each of the some 30,000 Americans with cystic fibrosis and their families may react, cope, develop, and adjust differently, it is crucial for social workers, psychologists, and nurses to have some theoretical basis from which to address their needs. Dr. John Rolland Family-Systems-Illness Model offers a theoretical framework particular to the varying needs of different families coping with illness (Rolland, 1994).

ASSESSING FAMILY ADJUSTMENT

The Family-Systems-Illness Model provides a useful, systemic view of family adaptation to serious illness in a child as a developmental process over time (Rolland, 1994, 12). The complexity of this model is particularly well suited to understanding the varying needs of individuals and families with cystic fibrosis. Beginning with a psychosocial typology of illness, the Family-Systems-Illness Model describes the intricacies of psychological and familial issues that interplay with a particular disorder's characteristics.

The typology conceptualizes patterns of onset, course, outcome, type and degree of incapacitation, and degree of uncertainty (Rolland, 1994, 23). It lays out a psychosocial understanding of illness in systems terms, including the expected pattern of practical and emotional demands of disorders over time, and looks at the timeline of a disease in its relevance to the related developmental tasks associated with different levels of the disease as it unfolds. In addition, the Family-Systems-Illness Model seeks to understand the values, beliefs, and multigenerational legacies that underlie health problems and care giving systems. Lastly, the model is sensitive to the undercurrent of anticipatory grief and separation that permeates all phases of individual and familial adaptation (Rolland, 1994, 29).

The model fits particularly well with assessing adjustment in a family with cystic fibrosis because it allows for variety in development and needs. Rolland addresses issues relating to CF as a slowly progressing, gradual, and unpredictable illness. The Family-Systems-Illness Model helps account for the particular psychosocial demands for individuals and families facing the effects of a perpetually symptomatic family member whose disability increases in a stepwise and progressive fashion. The model acknowledges the difficulty of continual and perpetual adjustment for children and families with CF, that family

members can never settle into any one phase because uncertain disease progression looms (Rolland, 1994, 24). But, this model also touches on the strengths and "goodness of fit" for particular families and particular illnesses. A family with stamina and flexibility can have great success in coping with the changing needs of a child with CF. Most importantly, the Family-Systems-Illness Model normalizes the complex, mutual interactions between the disorder, the patient, and the family (Rolland, 1994, 9).

Within a medical or hospital mental health setting, the social worker would have an important role to play in both assessment and supportive aid to the family caregiving system. Because social workers are trained to interpret, understand, and help families and individual clients understand the many interacting systems that can affect family adjustment, the social worker can be a lifeline to families attempting to navigate the many different levels of need and adjustment necessary to coping with and living with cystic fibrosis. Throughout the initial assessment and long-term treatment process, the social worker can help and advocate for families and their various needs.

When assessing a family caring for a child or adolescent with cystic fibrosis, a social worker needs to look at issues important for any family of a child with a chronic or terminal illness. For cystic fibrosis, it is important to ask about the meaning of the illness to that family: What are their individual expectations about health, daily living and life span? A social worker would need to explore the family's past strengths and deficits in dealing with losses and grief. It would be important to explore the flexibility or elasticity of familial roles, and family members' willingness to use outside resources when necessary. In addition, the health care professional would need to carefully assess a family's financial situation, with particular attention to health insurance coverage. Lastly, and perhaps most importantly, the social worker should make sure that a family and child with cystic fibrosis know about the positive outlook for treatment and life span while maintaining open communication about health and prognosis. One of the most valuable and life-saving things a family can do is find the strengths and abilities each member brings to coping with CF. Children, adolescents and young adults with cystic fibrosis are often described by others as having infectious laughs, a positive attitude, adaptability and great faith (CFF, 2002; Hollander, 1997; Lubich, 2002). By working to support those individual strengths, and by building and maintaining medical and social support networks, families and individuals with cystic fibrosis can begin to face the changeable course of this disease and can hopefully live full and meaningful lives. ■

REFERENCES

Cystic Fibrosis Foundation. (2002, April). *Facts about Cystic Fibrosis.* Retrieved April 20, 2002, from http://www.cff.org/facts.htm

CYSTIC-L. (2002, April). *CYSTIC-L: An email support group and newsletter for people with Cystic Fibrosis and their friends, families, and health-care providers.* Retrieved April 20, 2002, from http://www.cystic-l.com/handbook/

Desrosiers, C. (Director), Dufour, C., & Verville, L. (Producers). (1996). *Cystic fibrosis in adults* [Videotape]. Canada: Multimedia Group of Canada

Harris, A., & Super, M. (1995). Chapter 6: Psychology of cystic fibrosis. *Cystic fibrosis: The facts.* New York, N.Y.: Oxford University Press Inc.

Hollander, F. (Producer/Director). (1997). *Living with cystic fibrosis* [Videotape]. Sherborn, Mass.: Aquarius Healthcare Videos.

Lubich, L. (2002, April). *How to handle living with Cystic Fibrosis.* Retrieved April 20, 2002, from http://www.geocities.com/HotSprings/Villa/4210/

National Heart, Lung, & Blood Institute (NHLBI). (2002, April). *Facts about Cystic Fibrosis.* Retrieved April 20, 2002, from http://www.nhlbi.nih.gov/health/public/lung/other/cys_fib.pdf
Pulmonology Channel. (2002, April). Cystic Fibrosis lung treatment. Retrieved April 20, 2002, from http://www.pulmonarychannel.com/cf/treatment_lung.shtml

Rolland, J.S. (1994). *Families, illness, & disability: An integrative treatment model.* US: Basic Books.

Shapiro, B.L., & Heussner, R.C. (1991). *A parent's guide to cystic fibrosis*. Minneapolis, Minn.: University of Minnesota Press.

Silverstein, A., Silverstein, V., & Silverstein, R. (1994). *Cystic fibrosis.* New York, N.Y.: Franklin Watts.

ABOUT THE AUTHOR

GRETCHEN LANKA ALLEN is a second-year clinical student at SSA, whose main clinical interest is promoting family adjustment and coping with families, children and adolescents with chronic or terminal illness or disability. Gretchen is originally from northern California, went to Dartmouth College in Hanover, N.H., and has worked extensively with children with diabetes and other chronic illnesses. She also worked in outpatient psychiatry research at Massachusetts General Hospital for two years before attending graduate school. Last year, she was an intern at a public special education school in Skokie, Ill., and this year she is an intern with the Perinatal Family Support Center at Evanston (Ill.) Hospital.