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The Cystic Fibrosis Patient- A Nursing Challenge Met!

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What is cystic fibrosis?

UC San Diego Medical Center has a long history of caring for persons affected by cystic fibrosis. CF is an autosomal recessive genetic disease affecting approximately 30,000 adults and children in the United States. This disease results in excessively thick mucus production, which in turn affects multiple organ systems and leads to life threatening lung infections and severe gastrointestinal problems. Cystic fibrosis is progressive and eventually terminal. Millions of Americans carry a single CF gene, but are asymptomatic and typically not aware. A person with CF must inherit two CF genes, one from each parent, to produce the disease. Including an estimated 1 in 29 Caucasian Americans, CF is the most common, deadly, inherited disorder affecting Caucasians in the United States.

Diagnosis of CF

Most children with CF are diagnosed by age 2. Some persons, however, are not diagnosed until age 18 or older. These patients usually have a milder form of the disease. Genetic (blood) testing is now available to identify the genes resulting in CF.

Challenges in CF

Persons with CF face many challenges. Efforts to minimize flare ups must be performed throughout the day and include oral and nebulized medications. Airway clearance therapies may include the use of devices such as The Vest®, Acapella®, and exercise to loosen and expel accumulated mucus in the lungs. Routine therapies for adults usually require 4-6 hours daily when well and average 12-16 hours when ill.

In addition to the many complications faced by people with CF and the time intensive therapies involved, the financial burden is tremendous. Persons with CF are

frequently unable to work due to decreased pulmonary function and the time commitment necessary to complete the daily treatment regimen. Often cystic fibrosis is considered a preexisting condition, making insurance difficult to obtain. Recent health care reforms may alleviate some concern but are yet to materialize.

UC San Diego Adult CF Program

First established in the 1970's, the UC San Diego Cystic Fibrosis program cared for a small number of both children and adults with cystic fibrosis. Historically, CF had been considered a pediatric disease, primarily due to the fact that in years past individuals with CF rarely survived into adulthood. Improvements in technology, treatment, and medications have resulted in significant increases in median life expectancy. In the 1950's a child born with CF rarely survived through elementary school, in the 1980's this improved to an average of 25 years and today persons with cystic fibrosis have a median life expectancy of over 37 years (Cystic Fibrosis Foundation). In fact, within the last two years, adults affected by CF now total 47% of the disease population and will soon surpass children for the first time in history.

Much of the success in treatment and life expectancy for CF can be attributed not only to individual dedicated professionals but to the Cystic Fibrosis Foundation (CFF). Originally founded by a group of parents and families seeking to raise funds and seek a cure, the CFF has now grown to a large network of patients, families, and professionals. The CFF is now the accrediting body for the CF community and our accredited center which consists of both Rady Children's and UC San Diego.

As people with CF began to live



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longer, the UC San Diego CF Program quickly recognized that adults living with CF had unique healthcare needs and would be best served by establishing a separate Adult CF Program. The pediatric program moved to Rady Children's Hospital in the late 1990's and although now located in a different physical location, the two programs work closely together. The Adult Cystic Fibrosis Program was established in 1995 with the a patient population increasing to about 80 by 2005. Within the last 5 years the program has experienced tremendous growth, resulting in a current population of

Mucus blocks air sacs (alveoli) in the lungs



Mucus blocks pancreatic ducts



greater than 180 adults with CF that are cared for at UC San Diego.

Multidisciplinary Care

Long before it was in “vogue”, multidisciplinary care has been the delivery model of choice for the Cystic Fibrosis Program. A physician, nurse, and social worker have been integral members of the team for decades. In recent years the team has expanded and now includes two physicians, a nurse coordinator/program manager, registered nurse, dietitian, social worker, respiratory therapist and administrative assistant. Expanded members include audiology, PFT staff, research staff, medical assistant and pulmonary fellows. Team members are highly regarded for their expertise and individual contributions to achieving the highest quality care possible.

CF Nursing

At the heart of the multidisciplinary team is the Nurse Coordinator. The Nurse Coordinator is responsible for coordinating virtually all aspects of care for the cystic fibrosis population at UC San Diego. As one can imagine, a complicated disease such as cystic fibrosis requires skilled care coordination. The CF Nurse Coordinators provide outpatient case management, home IV antibiotic coordination, patient advocacy, patient and family education, staff education, research support, and more to people with cystic fibrosis.

A day in the life of the CF Nurse Coordinator is not for the faint of heart. The level of effort required daily to help maintain the health of people with CF is usually surprising to those unfamiliar with this population. Outpatient coordination includes clinic

preparation and follow-up, telephone triage, medication refills and prior authorizations, insurance assistance, maintenance of national databases, and much, much more. Medications prescribed for CF are unusually expensive. Obtaining approval for these routine CF therapies is challenging and often requires extensive paperwork and telephone calls to secure coverage. On average, 10% of the patient population is on home IV antibiotic therapy. Approximately 45% of the patient population lives 2 hours or more away from the medical center, creating significant challenges in the coordination of home health providers and nursing care. Although some of the daily requirements of outpatient care coordination may initially appear routine, they are critical to assist patients in maintaining their prescribed healthcare regimen. The CF Nurse’s role is essential to ensuring that this aspect of each patient’s care is well coordinated, allowing them the ability to simply live life to their fullest of their ability. The importance of even the most basic task cannot be underestimated.

The concept of nurse as patient advocate is not new. Nurses have historically advocated for their patients and it is not surprising that in a population such as CF that this quality is well developed. Just as persons with CF are taught from an early age to fight, fight, fight in order to survive, CF nurses are also tenacious. Whether it is for a needed medication, medical procedure, or expensive equipment, CF nurses quickly learn to fight their battles as patient advocates. Some patients have been let go from their jobs due to the expense to smaller employers’ insurance coverage premiums or

dropped from college classes due to repeated or prolonged hospitalization. Others have been denied, secondary to lack of insurance coverage, the ability to receive their care at our UCSD Adult CF Program, the only one of its kind in San Diego. Nurses may personally advocate or may educate patients and families in navigating the system and tools available to empower them to advocate for themselves.

CF Nurse Coordinators provide education to all members of the community involved in the care of persons with cystic fibrosis. Education is provided to patients and families regarding all aspects of the disease. Adults newly diagnosed with CF require extensive education on the basics of CF. As persons advance through different developmental stages, their educational needs change as well. CF Nurses not only identify needs as they arise but also anticipate needs before they emerge. Classes are provided to staff throughout the year including CF 101, provided through the new grad program. Nurse Coordinators present at state and national conferences regarding various aspects of CF. Education is an integral component of CF care. CF Nurses are life long learners, continually advancing their knowledge as the field of cystic fibrosis evolves.

As persons with CF live longer, healthcare needs become more and more complicated. The CF Nurse Coordinators provide support to patients and families as they are undergoing the decision making process for bilateral lung transplantation, awaiting lung transplant, and dealing with the multitude of concerns that arise as the disease progresses. Nurses actively participate in end of life and palliative care processes and support patients and families in a myriad of ways at this most difficult time.

The role of the CF Nurse

Coordinator continually evolves as the needs of patients and the program change. CF Nurses are passionate, knowledgeable professionals, dedicated to meeting every challenge, great and small, to allow persons with cystic fibrosis to thrive.