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# Cystic Fibrosis

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Cystic Fibrosis (CF) is an autosomal, recessive genetic disorder that affects approximately 40,000 children and adults in the United States and about 105,000 people worldwide (Cystic Fibrosis Foundation, n.d.). In people with CF, there is a defect in the CF transmembrane conductance regulator (CFTR) gene. There are many different mutations that a person can have, but the most common is Delta F508. CF affects the movement of chloride, a component of salt, to the cell surface causing the mucus to be thick and sticky. The thick mucus production can make people with CF more prone to lung infections since this mucus cannot be cleared as efficiently. It also often affects the pancreas, liver and gastrointestinal function, which can result in diabetes, liver disease, and malnutrition, leading to poor growth. CF also impacts the reproductive system, such as an absence of the vas deferens in men and infertility problems in women. Even though infection control concerns call for people with CF to distance themselves from one another, the CF community truly bands together for the greater good, helping all affected by CF. One of the ways they accomplish this is through the Cystic Fibrosis Foundation (CFF), a nonprofit organization founded in 1955 by a group of parents of children with CF. Since their founding, the CFF has partnered with the CF community to change the trajectory of CF and has contributed to

many advancements made in CF over the years (Cystic Fibrosis Foundation, n.d.). People with CF are often well informed regarding their disease. The CF Foundation partners with patients and their families, further encouraging those with CF to be actively involved in their care.

Cystic fibrosis used to be considered a childhood disease because many people with CF did not live to adulthood. However, due to medical advances and the recent development of CFTR modulator therapies such as Kalydeco® and Trikafta®, life expectancy for people with CF has increased significantly over the past decade. In 1955, children with CF were not expected to live long enough to attend elementary school. Through the dedicated efforts of the Cystic Fibrosis Foundation and people with CF and their families, new treatments and medications have been developed. CF is now a chronic disease, with people over 18 comprising more than half of those with CF in the United States (Cystic Fibrosis Foundation, n.d.). The median predicted life expectancy in 2020 for someone with CF born in 2017 was 46.2 years of age. About 75% of people with CF are diagnosed before age 2 and as more genes associated with CF are identified, some diagnoses occur well into adulthood (Cystic Fibrosis Foundation, n.d.). CF is a complex disease that affects most bodily systems and care in CF requires specialized knowledge. CF Foundation-accredited care centers offer the best care,



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UC San Diego Health Cystic Fibrosis Team

treatments, and support for those with CF.

UC San Diego Health Adult Cystic Fibrosis Program is one of the few CF Foundation accredited adult care centers in the region. Accreditation requires adherence to strict guidelines established by the Cystic Fibrosis Foundation (CFF). Accreditation of CF centers promotes standardization of care across centers and helps to build a comprehensive network dedicated to common goals—improving the lives of people with CF, development of new treatments, and ultimately, a cure for CF. The Cystic Fibrosis Foundation provides funding and accreditation to 130+ care centers across the US (Cystic Fibrosis Foundation, n.d.). The UCSDH program was established in 1996 by our current medical director, Dr. Douglas Conrad. At that time, there was also a part-time social worker and nurse working with the program. Our former program manager, Jessica Goggin, PhD, RN, MSN was also integral to the growth and development of the CF program. What started out as a small patient population (<50) and one MD/RN/SW team has developed over the years into a diverse, multidisciplinary team caring for approximately 270 people with CF and their families. UC

San Diego Health partners with Rady Children’s Hospital San Diego to form a comprehensive cystic fibrosis center. While most patients who receive specialized CF care at UC San Diego reside in the San Diego region, many travel from farther away, including Riverside County, Mexico, and Hawaii. In addition to adults with CF, the team provides clinical expertise and care coordination to people with other lung conditions such as complex bronchiectasis, primary ciliary dyskinesia, nontuberculous mycobacteria infections, and those with lung complications undergoing bone marrow transplant.

To provide the complex medical care needed by adult CF patients at UCSDH, the multidisciplinary team includes pulmonologists, nurse coordinators and LVN, social workers, a respiratory therapist, pharmacist, and registered dietitian. The program manager, scheduling and authorization coordinators, and research coordinators provide additional layer of support. Each member has developed expertise in the care and treatment of people with CF. The team ensures that patients’ needs are addressed taking the whole person into consideration. We aim to provide comprehensive care that

supports patients to achieve a high quality of life based on their individual goals and priorities. Through a multidisciplinary approach to care, patients can see several specialists in one visit. Some patients need IV antibiotic therapy frequently but may not need hospitalization. Our team of nurse coordinators and pharmacist help to ensure that patients’ care experiences in the home environment are comparable to the high-quality monitoring and treatment available in the hospital setting. They work diligently to deliver care in an efficient, timely, and safe manner by coordinating with home infusion pharmacies and home health nursing. The CF Team has developed a well-organized monitoring program and other clinicians and departments seek out their expertise. We also collaborate with other specialists within the organization such as the inpatient team, pulmonary function testing staff, audiologists, endocrinologists, gastroenterologists, otolaryngologists, and the lung transplant team.

UCSDH Adult CF Program conducts cutting edge research, a part of which is its participation in the CF Therapeutics Development Network (TDN). An integral component of this



network is the CF Foundation Patient Registry, established in 1966 which collects information on health status and outcomes of those with CF who have consented to participate (Cystic Fibrosis Foundation, n.d.). Collecting and analyzing information from the patient registry has made significant contributions in the development of clinical care guidelines, informing future research directions and driving quality improvement initiatives. One of the biggest challenges in the treatment of CF is antibiotic resistance. UC San Diego Health is helping to address this issue by actively participating and recruiting for research studies and offering other treatments for infection such as phage therapy. While the concept of phage therapy is not new, UCSDH is one of the few institutions in the US equipped to provide this treatment. “As a premier academic medical center, UC San Diego Health provides specialized education in cystic fibrosis through our medical school, allied health training programs, and our Pulmonary and Critical Care Medicine Fellowship Program” (UC San Diego Health, n.d.).

People with CF are a special group. It is with great privilege that our team is involved in taking care of this unique patient population. Many have required multiple hospitalizations for pulmonary exacerbations and other ailments over the years. Living with CF is not easy, in addition to physical and emotional symptoms, treatment is time consuming. Treatment burden for people with CF is high, with some taking upwards of 50 pills per day, multiple respiratory treatments to help thin their mucus and open their airways, as well as inhaled antibiotic therapy to suppress the growth of bacteria in their lungs. People with CF also use airway clearance devices to help mobilize secretions so that they can expel the thick mucus from their lungs. In total, people with CF spend about 4-6 hours daily taking care of their CF needs, which can increase to 8+ hours if they are experiencing an exacerbation. That is equivalent to a full-time job! What is most amazing about this population is that despite

this high treatment burden, people with CF are achieving great things. I will never forget one of our patients, recalling how when she was a child, she never considered having children of her own someday, going to college, or living into adulthood because of her CF. But now, those things are possible! It is so rewarding to watch them attain a goal or milestone that they may have thought at one time was impossible.

As mentioned earlier, the CF Community exhibits a level of solidarity that is not always apparent in other chronic disease populations. Many people with CF and their families are very active in giving back to the CF community whether it is through their professional work, fundraising efforts, volunteering with the foundation, educating the public, or participation in research. We have witnessed firsthand how selfless people with CF can be, for example, participating in a study that may not benefit them directly, knowing that this may help improve the quality of life and future care for others with CF. They remain a close-knit community that seeks out ways to come together and provide support for one another. In 2020, the CFF embarked on a journey, “Path to a Cure”, which targets the underlying causes of CF and aims to further the development of treatments for all people with CF. As their motto states, “Make CF Stand for Cure Found”. If the past 50 years are any indication of success, together, the CF community and the CF Foundation will make this a reality. The UC San Diego Health Adult Cystic Fibrosis team are honored to provide care to the people with CF and their families in our community.

## REFERENCES:

- UC San Diego Health. (n.d.). Adult cystic fibrosis program [Brochure].
- Cystic Fibrosis Foundation. (n.d.). Guide: About cystic fibrosis. Retrieved January 25, 2023, from <https://cff.org/intro-cf/about-cystic-fibrosis>
- To learn more about Cystic Fibrosis, please visit the Cystic Fibrosis Foundation website at [www.cff.org](http://www.cff.org)