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I am a 2024 graduate from the Columbia School of Social Work on the Integrated Practice and Programming track and specializing in the study of health, mental health, and disabilities. I am currently living in Columbus, Ohio, and excited to see where this field takes me next.

INSPIRATION FOR ARTICLE

My practicum is at Nationwide Children's Hospital in Columbus, Ohio, where I am a social work intern in the division of pulmonary medicine. Nationwide Children's is home to one of the highest ranked Cystic Fibrosis Centers in the country and serves around 500 people with CF in Ohio, West Virginia, and Kentucky. Through this placement, I have worked with many people of all ages who have CF, and have recognized the psychosocial impact that new, revolutionary medications have had on the lives of these individuals. I have been so inspired by the resilience and strength of those that I have worked with. As there is limited research on this topic, I hope this paper becomes part of a larger conversation on how we can support the unique experiences of people with CF.

ABSTRACT

Cystic fibrosis (CF) is a chronic, progressive, genetic, and life-limiting lung disease that impacts approximately 105,000 individuals globally, including 40,000 individuals in the United States. In 2019, a revolutionary new drug, elexacaftor/ivacaftor/tezacaftor (ETI), was approved to manage some of the major symptoms of CF and dramatically increase the lifespan of people with cystic fibrosis (pwCF). Many individuals with CF cannot work full time and require Medicaid, Supplemental Security Income (SSI), Social Security Disability Insurance (SSDI), and other financial assistance programs to pay for treatments and medical expenses. Yet in recent years, pwCF who are on ETI have been increasingly losing benefits. A possible explanation for this is the effectiveness of ETI in improving lung function, creating the perception that pwCF are no longer disabled by their condition. Even with this "miracle" drug, pwCF continue to experience significant complications and vulnerabilities to their physical and mental health as well as limitations on daily living and employment. The compounded vulnerabilities these individuals experience leave them without a safety net. Social Security policies for pwCF require revisions to prevent further biopsychosocial damage to this population. Two policies will be recommended: leniency in redetermination, and CF education for those who make determination decisions.

INTRODUCTION TO CYSTIC FIBROSIS CARE

"What do you do when you've lived your whole life diagnosed with a terminal illness and then are suddenly diagnosed with new health? For me, it was not a whimsical or magical moment."

Luisa Palazola (BioNews Staff, 2021, para. 4).

Cystic fibrosis (CF) is a chronic, progressive, and life-limiting multisystemic genetic disorder, impacting approximately 105,000 individuals globally, including 40,000 individuals in the United States (Cystic Fibrosis Foundation [CFF], n.d.-a). Impacting the lungs, pancreas, gastrointestinal (GI) system, reproductive system, and other organs, CF presents differently from person to person but commonly causes coughing, difficulty breathing, frequent lung and sinus infections, and poor weight gain. Managing a demanding condition like CF brings significant social, physical, and psychological challenges. Socially, it is an isolating disease, as people with CF (pwCF) are not allowed to share spaces due to their susceptibility to bacterial infections and the risk of spreading infections to other pwCF. This, in turn, prevents community building through means such as in-person group therapy or sharing classes in schools. Physically, pwCF typically need to spend several hours per day managing their condition, whether through administering medications and lung therapies, preparing special diets, or attending doctors' appointments. Mentally, this heavy treatment burden can take a psychological toll, putting pwCF at increased risk for developing a mental illness, while the required isolation creates additional psychosocial challenges (Vines et al., 2018; Bathgate et al., 2022).

In 2019, a revolutionary new drug, elexacaftor/ivacaftor/tezacaftor (ETI), which targets the genetic cause of CF, was approved by the Food and Drug Administration (FDA) under the brand name, Trikafta (U.S. Food and Drug Administration [FDA], 2019). CF is caused by mutations in a gene called the cystic fibrosis transmembrane conductance regulator

(CFTR). All people possess two copies of the CFTR gene, but for pwCF these genes are mutated, leading to dysfunctional CFTR proteins that interfere with proper cell hydration, causing the mucus covering these cells to thicken (CFF, n.d.-a). ETI combines three types of CFTR modulators to help correct these protein defects (FDA, 2019; CFF, n.d.-d).

This new medication has dramatically increased life expectancy for pwCF. Prior to the emergence of ETI and similar therapies, the projected lifespan for pwCF was approximately 37 years old (Lopez et al., 2023). If ETI is started between the ages of 12 and 17 and taken as directed by a patient with good lung health, ETI will nearly double one's projected lifespan to 82.5 years old (Lopez et al., 2023; FDA, 2019; Ladores & Polen, 2021). In addition to life expectancy, ETI also increases pwCF's ability to get pregnant and improves pulmonary function to the extent that patients are able to withdraw from lung transplant waitlists (Ladores & Polen, 2021).

An estimated 90% of pwCF have the F508del gene mutation that makes them eligible to take ETI, while around 10% of other pwCF have different gene mutations that make ETI and other modulator therapies ineffective (FDA, 2019). ETI and other modulator therapies are treatments, not cures. This means that despite these remarkable medical advances, CF remains a complex and often disabling condition requiring frequent medical checks and a high level of medication and diet adherence while continuing to pose an increased risk of lung infections and other physical and mental health issues. As a result, many individuals with CF require Medicaid, Supplemental Security Income (SSI), Social Security Disability Insurance (SSDI), and other financial assistance programs to maintain a stable foundation (CFF, n.d.-b; CFF, n.d.-c). Medicaid is a critical resource and can help cover ongoing costs of treatment for pwCF, including ETI's hefty price tag of more than \$300,000 per year in the United States (McConnell et al., 2020; Wexler, 2023). Yet Medicaid does not cover anything beyond what is considered medical care (CFF, n.d.-b). This means SSI and SSDI are crucial supports for individuals

disabled by CF-related conditions, particularly those who remain unable to work (CFF, n.d.-c).

As a social work intern at a CF center at a hospital in Ohio, I have been exposed to the countless biopsychosocial struggles of pwCF and have had many questions about what can be done to support this population. One trend identified by patients at this hospital in the last few years has been a significant increase of pwCF on ETI losing their Social Security Administration (SSA) benefits. The multidisciplinary hospital team attributes this to ETI's effectiveness in improving lung function so much that on the surface, pwCF appear to be no longer disabled by the disease.

This trend raises concerns since, even while taking this "miracle" drug, pwCF continue to experience considerable physical and psychological challenges and vulnerabilities and are frequently still admitted to hospitals for surgeries, pulmonary exacerbations, lung infections, GI issues, and other complications. Since ETI is not a cure, adults with years of lung scarring may only see minor improvements in lung function or simply may not see further damage (K. Pasley, personal communication, March 22, 2024). For many on ETI, they are not thriving, just surviving.

While official data on rates of removal of financial benefits are limited, the increase in first-hand accounts is alarming, and CF social workers and other professionals who have observed the financial consequences of losing this support have struggled to identify resources to help pwCF (CBS News, 2019). This population has struggled with this disease for their entire lives, and because CF was previously considered a "death sentence," they were not adequately prepared for such an increase in life expectancy without financial support. Their previous prognoses meant they often did not invest in their education, personal relationships, financial investments, and careers, and as a coping strategy "often 'do things now rather than [*sic*] later' as a strategy to experience their limited and precious lifetime to the fullest" (Moola, 2019, p. 360). Now that the SSA appears to be increasingly

removing benefits for those on ETI, those impacted are forced to seek employment. However, due to the nature of the disease, the demands of daily treatment, and having limited lung function, many are physically unable to attend school or work full time. The compounded vulnerabilities these individuals continue to experience make the loss of a safety net even more challenging.

Social Security policies pertaining to pwCF need to be revised to prevent further biopsychosocial damage to this population. Two case studies from Ohio will be shared to provide further insight into the struggles these individuals face as they transition to this new era with ETI. Policy recommendations will then be discussed, including leniency in SSI and SSDI redetermination and CF education for those who make determination decisions. Lastly, social work implications will be discussed, including disparities and best practices for supporting this population with the life changes that accompany ETI therapy.

CASE STUDIES FROM OHIO

One individual seen in the CF clinic at a hospital in Ohio exemplifies why vulnerable pwCF benefit from Social Security coverage even as their health improves (K. Pasley, personal communication, November 11, 2023). This individual is a 24-year-old who grew up thinking he would die young due to this disease. He struggled to get an education, never learned how to adequately take care of himself, and was unable to generally thrive. He was 22 years old when ETI became available. After ETI dramatically improved his respiratory health, he lost Social Security benefits. He has not been able to maintain a steady job as his lifelong illness prevented him from learning critical skills, and, despite his improved lung function, he has struggled to regularly attend work hours due to being sick, often taking leaves of absence. As a result, his financial situation remains unstable and he has continued to rely on the financial safety net of Social Security.

Another individual seen in the CF clinic, a 50-year-old male with CF, was able to work for years before becoming disabled by his illness. He

had never experienced any interruptions to receiving SSDI benefits until ETI became available. He was the first person in this CF clinic to raise concerns about what might happen to his SSDI if he became healthier when he took ETI. At 50 years old, he was accustomed to his quality of life with SSDI and was not interested in trying to re-enter the workforce due to his age and health. He said that he would even choose to stay off of ETI in order to keep his benefits.

These stories have become increasingly common. In 2019, an attorney who runs the Cystic Fibrosis Legal Hotline noted,

We've seen a five-times increase in the number of people with cystic fibrosis that have been reviewed in the past 18 months. And we think that Social Security is targeting young people with chronic illness in an effort to reduce the number of people getting benefits (CBS News, 2019, 2:45).

Given this pattern, many pwCF are understandably concerned about choosing between their health, their job, and Social Security benefits.

RATIONALE FOR ACTION

BENEFITS OF SOCIAL SECURITY AS A SAFETY NET

Financial benefits, including SSI and SSDI, are an important lifeline for pwCF. SSI redetermination, which evaluates financial resources and living arrangements, occurs every 1 to 6 years, and Continuing Disability Review (CDR), which evaluates the medical condition of SSI and SSDI recipients, occurs every 1 to 3 years (Social Security Administration [SSA], 2024a; SSA, 2024b; SSA, n.d.-c). If one has a medical condition that is not expected to improve, such as CF, SSDI redetermination will occur every five to seven years, though usually seven years for pwCF (SSA, 2024b; CFF, n.d.-c). Redetermination considers whether one's health has improved over 12 months; financial benefits are rescinded if one's health has improved to a point where one can work (CFF, n.d.-c).

The SSA also discontinues benefits if someone earns more than \$1,550 per month (SSA, 2024c). If a person with CF earns more than the

maximum limit, even if their health is still precarious, they lose benefits, and if their illness later prevents them from working, they become financially vulnerable. A social and financial safety net is therefore vital for improving employment and housing stability, maintaining continuity of health care, and avoiding added financial stress (Crane et al., 2019). SSI and SSDI programs provide this much-needed safety net while the loss of benefits puts an already vulnerable population at even greater risk.

MENTAL HEALTH CONCERNS FOR PWCF TRANSITIONING TO A LONGER LIFE EXPECTANCY

A number of studies have found ETI to impact mental health and social functioning in pwCF in complicated and even paradoxical ways. Several studies found, for example, that improved pulmonary health was correlated with improved mental health (Hjelm et al., 2023). However, some studies looking at the effects of ETI found that, while the medication improved physical symptoms, ETI was correlated with worsening mental health symptoms, sleep issues, and increased rates of changing or initiating psychiatric medication (Zhang et al., 2022; Bathgate et al., 2022). Other studies found paradoxical results on ETI's impact on mental health (McCoy et al., 2023; Piehler et al., 2023; Zhang et al., 2022), and some found no statistically significant improvements in anxiety scores, depression scores, emotional functioning, or perception of body image (Zhang et al., 2022; Finlay et al., 2021).

One explanation for this lack of improvement or worsening of mental health symptoms may be the sudden change in longevity and quality of life. Prior to these new modulator therapies, CF was known as a "child killer," and many pwCF were told from a young age they would have a limited lifespan (Kempner, 2022). One individual reflected, "I was just living day-to-day instead of planning for the future," (Kempner, 2022, p.3) while another stated she had "never pictured a future for herself beyond the next five years" (Kempner, 2022, p.1). As a result, many pwCF who grew up without these new modulators have not invested in their life, health, education, or careers. They may not have created

long-term goals nor developed independence or coping skills. This is exacerbated by the fact that many PwCF experience high rates of comorbid mental health diagnoses, including depression, anxiety, obsessive-compulsive disorder (OCD), medical traumatic stress (MTS), attention-deficit hyperactivity disorder (ADHD), autism spectrum disorder (ASD), oppositional defiant disorder (ODD), and disordered eating (Bathgate et al., 2022; Lord et al., 2022; Guta et al., 2021). Having grown up with a terminal illness mindset, many are now struggling to establish long-term goals as they adjust to a sudden and unexpected increase in life expectancy. Additionally, the risk of early mortality did little to motivate pwCF to establish positive health habits such as routines around eating, sleeping, and CF medical care.

This adjustment is also compounded by the fact that, even with ETI, PwCF often have difficulty completing instrumental activities of daily living (IADLs), which include managing medications, money, and transportation. This makes full-time work challenging, leading to lower employment rates and, consequently, lower rates of employment-based insurance (Callahan & Cooper, 2007). PwCF who do work often end up needing to take long leaves of absence due to susceptibility to illness, CF exacerbations, and hospitalizations. As a result, they rely on Social Security benefits to help cover living and medical expenses. A 2019 study on the experiences of Ohioans with co-occurring chronic health conditions and mental health impairments found many experienced financial hardships such as unemployment and/or financial instability and, in turn, had difficulties paying for food, rent, and other necessities (Crane et al., 2019).

According to one individual with CF who started ETI as a young adult, "to fathom what life would look like with stable health was incomprehensible to someone who never had that" (BioNews Staff, 2021, para 3). One study found that an individual who began ETI "encountered unanticipated internal turmoil" due to this change in life expectancy and no longer being seriously chronically ill (Ladores and Polen, 2021, p. 2). This individual elaborated that this transition period caused them anxiety as they thought that ETI would stop working: "I'm

so nervous because I feel so good and I'm not used to living this way, and I'm so afraid I'm gonna lose it" (Ladores and Polen, 2021, p. 3).

The Cystic Fibrosis Foundation recommends that pwCF ages 6 and older be seen by an accredited CF clinic at least 4 times per year (CFF, n.d.-e). There are more than 130 accredited CF clinics throughout the United States, each with a multidisciplinary team of doctors, nurses, nutritionists, respiratory therapists, social workers, and psychologists to provide comprehensive care for CF patients. These appointments require patience through a lengthy process of medical tests, scans, blood tests, and pulmonary function tests (Finlay et al., 2021). Social workers on these teams play an important role in identifying psychosocial and transitional needs of pwCF, meeting with patients yearly to conduct psychosocial reviews and assist with resource navigation and support (Finlay et al., 2021). Now, with these new modulator therapies, social workers are faced with the additional challenge of supporting a CF population as they adjust to the prospect of a longer life than they ever anticipated.

PROPOSED POLICY OPTIONS

To improve the livelihoods of pwCF, two policies are proposed in the state of Ohio which could be adapted across the country based on each state's specific SSA policies. The first is a higher level of leniency when redetermining eligibility for social security (SSI/SSDI). The second is a CF-specific training for consultative examination (CE) providers who complete SSDI evaluations and redeterminations. Currently, SSI/SSDI recipients who disagree with a reconsideration determination can request a hearing to appeal the decision (SSA, n.d.-a). However, this puts an undue burden on pwCF who may be physically unable to file the appeal or attend the hearing due to their condition and struggles with ADLs.

POLICY #1 - LENIENCY

A standard that allows for more leniency would help protect this vulnerable population from losing SSI or SSDI benefits simply because

they have been more physically healthy since beginning ETI or other modulators. As seen in research studies, improved physical health when taking a modulator does not necessarily mean one's mental health or ability to work will improve (McCoy et al., 2023; Bathgate et al., 2022). As this population transitions to having longer lives, a financial safety net would alleviate stress by providing stable resources to be used for basic necessities.

To implement this, the SSA would alter the qualifying definitions of "severity" and "frequency" of CF symptoms to make the removal of benefits a more medically-informed and thorough process. This change would occur in the SSA's evaluation manual at the state and local SSA offices in Ohio. The SSA does not publicly release eligibility criteria and only provides a "Listing of Impairments," which broadly mentions that the SSA will consider past work experience, severity of medical conditions, age, education, and work skills (SSA, n.d.-a). Due to this ambiguity in criteria, approval varies by each CE evaluator. To improve this system, eligibility requirements could be amended to include a section for pwCF that takes into account their age when beginning a modulator therapy, long-term goal planning, pulmonary function test levels, and other relevant medical complications. These factors would provide a more comprehensive picture of one's health and how they are coping with the transition to taking a modulator and having an extended lifespan. When considering job history, the criteria could be changed from "ability/inability" to work to "stability/instability" in work. This subtle yet significant change in language would expand eligibility to include those who experience leaves of absence due to CF-related medical issues. This more holistic framing better addresses the complexity of this disease and the careful management it requires.

POLICY #2 - TRAINING

In each state, disability claims are reviewed by CE providers, made up of physicians, psychologists, and other health professionals recruited by the SSA's professional relations officers (SSA, n.d.-b). Although these providers undergo extensive training, they are not experts in

every disease and its respective challenges. Without training in CF specifically, they are unlikely to recognize that, although on paper the health improvements from ETI therapy may give the impression that someone with CF can work full time, the mental and physical effects of their illness often remain debilitating. A second policy recommendation, therefore, is a one-time CF-specific training for CE providers to help them understand the nuances of this complex, lifelong disease and how important SSDI is for someone trying to manage it.

To implement this second recommendation, a committee from the Cystic Fibrosis Foundation (CFF) would select medical professionals in Ohio to provide training to the CE providers who process SSDI determinations and redeterminations. These one-time training sessions would be an hour long to remain cost-effective and would include subject matter experts (SMEs) who work with pwCF. CE providers who receive this training would be certified in CF redetermination and be primarily responsible for reviewing CF cases. The end goal would be to help evaluators make more informed decisions. The CFF would work with the SSA to continually monitor and review SSDI cases to ensure that pwCF are receiving proper support.

Allowing the most vulnerable pwCF to remain on Social Security would reduce costly hospitalizations and additional medical care. Someone who has their SSI/SSDI revoked will be forced to work more, putting them at increased risk of stress and infection. The safety net of Social Security allows them to work as much as they can within their own limitations, helping to reduce that risk.

SOCIAL WORK IMPLICATIONS

Social workers have continually played an important role in providing support to pwCF, and now with ETI, they must rise to the additional challenge of helping this population navigate the adjustment to a longer lifespan. One important way social workers can do this is by providing career counseling to pwCF who have a newly extended life expectancy and did not anticipate having to plan for a long-term career.

Social workers follow the guiding principles of competence, challenging social injustice, and promoting the dignity and worth of all people as they work to address social problems such as health inequity (National Association of Social Workers, 2021). As such, there is an important role for them to play in confronting racial and economic disparities among pwCF as treatments and technology improve. Because of the health disparities caused by structural racism, people of color with CF experience: overall lower lung function, nearly doubled rates of death before the age of 18, delayed diagnosis, higher rates of pulmonary infections, limited access to care, lower access to lung transplants, lower rates of health literacy, and are less likely to be represented in clinical research (Zampoli, 2023; CFF, 2020).

In terms of economic disparities, many low-income countries have less access to modulator therapies; in fact, only an estimated 12% of pwCF globally have access to CFTR modulators, like ETI (Guo et al., 2022, as cited in Zampoli, 2023). Low to middle-income countries in Central America, South America, the Middle East, South Asia, and Southern Africa have not been prioritized for negotiating contracts for accessing ETI (Zampoli, 2023).

While the U.S. does have contracts for manufacturing and distributing ETI, the U.S. government does not negotiate prices with drug manufacturers, leading to exorbitant costs (Martins, 2020; Ginsburg & Lieberman, 2021). The yearly \$300,000 list price for ETI is not feasible for the vast majority, leaving pwCF to rely on crucial government-funded insurance or employment-based insurance to afford this life-saving treatment. In addition, the multidisciplinary team at the CF center where I intern has noticed that several insurances are no longer covering ETI, creating an even greater need for these cost-assistance programs.

Social workers must work to lessen these disparities by centering the experiences of disproportionately impacted populations, providing education to care teams, and advocating for healthcare equity.

Finally, it is important for social workers to continue studying the unique psychosocial needs of this population. The current best measure of

psychosocial functioning for pwCF is the Cystic Fibrosis Questionnaire-Revised (CFQ-R), which includes assessment questions for physical health as well as social-emotional health. Yet this does not include questions on the effects of taking ETI. Social workers can advocate for questions on the impacts of this transition to ETI to be included in the assessment, which should then be used in clinical trials for any new CFTR modulator therapies. Accounting for these psychosocial factors can provide a more comprehensive picture of how the whole person is affected, moving beyond a narrow physiological perspective. Lastly, social workers should make sure to facilitate transparent and sensitive conversations with pwCF about potential and unanticipated impacts these life-altering medications may have (Ladores and Polen, 2021).

CONCLUSION

There are many unknowns about the quality of life for young people with CF who will grow up with these new medications. It is hoped that they will adjust well, need fewer safety nets, and plan and achieve long-term goals. However, it is clear that the current CF cohort who are suddenly facing a longer lifespan will need additional support. Social Security, including SSI and SSDI, has long provided a stable foundation for these individuals and should not be prematurely taken away. Advocates, such as social workers, should appeal to the SSA to be more lenient in redetermining eligibility and promote and provide CF-specific training to the CE providers who are assessing these cases. If this vulnerable population continues to lose benefits, it will have detrimental impacts on their physical and mental health and their overall quality of life. Because the successful increase in longevity from ETI treatment had the unintended consequence of creating new mental health challenges, the impacts of the stressors, real-life complications, and financial burdens associated with this drug should be studied further. Additionally, more research should be prioritized to understand racial and economic disparities of access to care for pwCF. While ETI has been revolutionary in improving the lives of people with cystic fibrosis, more work at the policy level must be done to show these individuals that their health and success matters.

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