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Cover Page Footnote

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Original parts: Aging and reckoning with cystic fibrosis related kidney disease

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Abstract

Kidney disease increasingly impacts people with cystic fibrosis (CF) as adult patients continue to survive longer. Yet the literature on CF related kidney disease focuses little on amplifying the voices of people aging with the condition. This article presents perspectives on CF related kidney disease from a medical sociologist who is themselves managing these issues. It (1) gives an overview of relevant literature and trends in epidemiological data on kidney disease and CF, (2) details the author's own process of adjusting to progressive changes in renal function, and (3) outlines opportunities for clinicians to make a positive impact for patients by centering their voices.

Keywords

Cystic fibrosis, kidney disease, aging, illness management, patient experience

Introduction

My kidneys and I recently celebrated 34 years of life—a milestone that might have seemed unrealistic had I been born in a different time. It seemed out of reach for me as well until I began getting coordinated care for my cystic fibrosis (CF), a disease that has likewise spent all 34 years with me but only received proper management well into my adult years. CF is a genetic disease in which the body cannot produce, transport, and/or use a specific type of protein that helps electrolytes get across cell membranes¹. It remains the most common fatal genetic disease among people of European heritage². Surviving and beginning to thrive with my CF has thus involved reckoning with many things beyond my own mortality as a general construct. Like any other patient, I constantly engage in illness management—the process of adjusting to changes in my health and allowing them to become part of my sense of normalcy³. And like any other scientist, I generally begin my processes of reckoning by examining data. In this narrative, I use my experiences to (1) give an overview of scholarship related to CF and kidney disease, (2) detail my experiences managing CF and kidney disease, and (3) outline ways clinicians can better serve patients by amplifying our voices.

An Overview of Scholarship on CF and Kidney Disease

Throughout my 34 years and counting, some trends in CF survival data have remained remarkably similar. Over 80 percent of people with CF still die from respiratory failure, even if they live for many decades before their lungs eventually succumb to the ravages of the disease⁴. That

leaves the other 20 percent of us who will likely still die from our CF, but from impacts to other organ systems as opposed to steep declines in lung function⁵. Kidney disease increasingly ranks among the leading causes of death for adults with CF⁶. This represents a change in the data from the early 1980s when I was born. My nephrologist here in Orlando commented that the learning curve for physicians has been just as steep as it has for patients with respect to chronic kidney disease, as only recently did people with CF begin surviving long enough in sufficient numbers to present a substantial case population.

Kidney disease is not a universal experience among people with CF, in part because so many patients continue to experience respiratory failure. The unique form of diabetes associated with CF can also harm the kidneys, but is observed in under 50 percent of the total disease population⁷. Likewise, not everyone who does develop CF related diabetes experiences kidney dysfunction as a result⁸. However, every person living with CF does experience elevated risk for renal impairment, especially as they grow older⁹. Several factors can contribute to kidney disease in people aging with CF. One prominent factor is obstruction of the glomeruli—the small ducts inside of the kidneys that transport fluids—with hardened pieces of mucus¹⁰. While I return to other pieces of CF or kidney disease where relevant later in the piece, these examples represent the overall picture of the intersection of these conditions in the existing scholarship. With this background in mind, I now shift to my own experiences before drawing on those experiences to make suggestions for clinicians in closing.

My Experience with CF and Kidney Disease

Years before my pulmonologist would ask how all my doctors in New Jersey never recognized these “casts” in my urine for what they were, I would stand beneath the lighting fixture in my parents’ kitchen as we all peered at a jam jar full of my urine. My mother and father, with PhDs in neuroscience and cell biology respectively and a collective four decades of experience as medical educators, were just as mystified as I was by the glowing white threads that floated in the liquid. Although we all spared a moment to appreciate the humor of collecting a urine specimen in a dainty little gingham-topped jar, there was also a sense of foreboding. Two years prior, I had been admitted to intensive care with a tentative diagnosis of renal failure.

Further testing—including the singularly peculiar experience of collecting all of my urine and feces for 24 hours while trying not to trip over my intravenous pole or drop my heart monitor into the toilet—revealed that my kidneys were stressed but responding as best they could to problems with other systems. The next time I was able to speak to my mother, who had frantically offered to give me one of her kidneys as my father haggled with airline ticket agents, I told her to keep her organs for the time being. With a serum potassium of 2.1 milliequivalents per liter that refused to budge even after oral supplements were introduced on top of the IV, I found humor where I could. But the laughter faded quickly as it became clear that the specter of kidney disease had long since become reality.

I spent years shuttling around to various nephrologists, first in central New Jersey and then in northern Florida as my increasing intolerance to cold drove me southward. Raynaud’s syndrome manifested early—a consequence of the vascular damage increasingly seen in adults with CF as survival increases¹¹. I wound up in Orlando, where care options for adults with CF are markedly better, through a combination of persistence and serendipity when FSU College of Medicine offered me a transfer to the Regional Campus here in exchange for my commitment to build out our community partnerships for aging research in the area. I enthusiastically accepted what amounted to a dream come true with no small sense of the irony inherent in it—someone with what remains a decidedly terminal illness leading research efforts centered around healthy aging. Yet that became an important milestone in my adaptation to living with the impacts of my CF, which at that point had been diagnosed conclusively, initiating a good deal of scrambling by myself and a vast team of clinicians to compensate for lost time.

Indeed, one of the other common causes of kidney damage in people with CF contributed strongly to the conversion of my diagnosis from a lifelong suspicion to a

formal pronouncement. I moved down to Orlando with a raging bacterial infection that had spread through my entire renal and urinary system. Such infections often spiral out of control in people whose CF is inadequately managed, as mine was for the better part of three decades. Paradoxically, some of the more aggressive drug therapies used for CF can also make the kidneys more vulnerable to bacterial colonization¹². I fell victim to this truism as well when ranitidine, a medication intended to reduce my gastrointestinal symptoms, aggravated my kidney issues so much that my entire lower body broke out in vasculitis rashes. A quick literature search revealed that vascular complications associated with kidney inflammation tend to occur more often in CF patients with substantial autoimmunity¹³ as already documented in my own medical records. I showed up at my primary care doctor’s office in Tallahassee, passed him a series of pictures on my phone, and said “I have Henoch-Schoenlein purpura”. Although this was far from a good day, it paved the way for much better ones to follow.

Months later, my new nephrologist would gently diagnose me with chronic kidney disease—“but only Stage 1, we still have plenty of time”—and send me off for a comprehensive renal and urinary ultrasound. While I waited for the results, I spent a lot of time thinking back to that day in the hospital when I told my mother to keep her kidneys, and all the times we had argued since about whether or not I should list for a transplant if my own failed. I thought about my pulmonologist’s confidence that by a combination of atypical genetics and hard work, I might survive long enough for my CF to become largely irrelevant for my life expectancy. Combing through endless arrays of literature and data, I concluded that although this was an outcome I very much wanted for myself and my family, getting there might require difficult decisions about my willingness to consider organ transplantation.

This meant that I needed types of data that are often more difficult to find in research journals—those that amplify patient voices and experiences¹⁴. I knew plenty of people with double lung transplants, something that had never been on the table for me once inhaled corticosteroids had rescued my pulmonary function¹⁵. As I got more involved in the CF community online, I also came to know several people who had undergone successful kidney transplants. Some of them even had stories like mine—relatively classic phenotypic cases of CF that fell through the cracks in a broken health care system. My first chloride sweat tests, once the gold standard for diagnosing CF, were performed at a scant few years of age. Insufficient sweat meant no conclusive diagnosis despite concerning results, and I floundered into adulthood with a halfway managed disease playing havoc with my body. I lost upwards of 40 pounds from my slender frame that I would never find again. I lived with chronic mycobacterial pneumonia for

most of my 20s. My kidneys screamed with pain daily. My legs swelled, the skin shining and painful with edema; my eyes bulged from my skull. I began to think about dialysis not as a question of “if” but rather one of “when”.

Yet things slowly began to improve. Some of the news I received was bad, if not unexpected—the kidney disease diagnosis, the evidence of thickened airways, the near-total failure of my pancreas to produce digestive enzymes anymore. Other pieces of news were better—airways only mildly thickened, lung function good and increasing, no diabetes despite the exocrine pancreatic insufficiency, kidney disease present but not progressing. I learned how to manage my kidney health better—not drinking inordinate quantities of water, managing my thirst instead with a combination of good hydration practices and mucus-thinning drugs, getting my medications adjusted to avoid adverse reactions, starting on special vitamin supplements for people with CF, staying away from anything that could damage the blood vessels further.

As I felt the seeds of empowerment taking root, I thought about the stories I had heard from friends online—people living happily with their transplanted kidneys, free from dialysis and the misery of waiting between sessions. I thought doubly hard about my mantra from all those years—“I’ll manage”—and wondered if perhaps that was an appropriate way of thinking now that I was no longer dying of a mysterious disease but rather living with a reasonably well understood one. I moved the goalpost accordingly—to be the oldest living person with CF, botched diagnosis and fragmented care be damned. I had learned that my genetics were not only atypical, but also likely to mean that I only had problems using the CF transmembrane conductance regulator protein rather than producing or transporting it, which likely meant my prognosis was better than those of many of my peers.

When the results from my renal ultrasound came back, I thought about those too. My nephrologist gave me a hopeful smile for the first time, saying that there was no severe damage to the tubules or other structures in my kidneys—just widespread inflammation and some light scarring. And as we continued to monitor my renal bloodwork and urine output, he said that my kidney disease did not appear to be progressing at a rapid rate. I contemplated the possibility of finishing out this life with all of my original parts, save the multiple teeth I had already lost and the gum tissue that had to be surgically reconstructed after its destruction by years of chronic infections. I still do not know if that hope will become reality for me, but I realized recently that I have come to a place in my life and health care where I can accept either alternative. I shared that with my parents in a lengthy series of text messages as if it were a small matter. Yet they knew, as I did, the seismic shift it represented in my thinking about the future.

Advice for Clinicians Serving Patients with CF and Kidney Disease

I share my experience now to highlight the importance of centering patient voices in care for kidney disease, whether related to CF or otherwise. Many people resist even discussing organ transplantation as an option because they have not gotten the opportunity to hear from recipients of successful donations¹⁶. Transplant can seem like an incredibly fatalistic prospect, especially for people who already know they are living with terminal diseases. The fact that your condition may take a very long time to kill you does not, I have learned, take away the fear that comes with thinking about potential kidney failure. I always said that I was not afraid to die, but rather afraid to suffer; that statement remains true for me today. I also still get regular tastes of the suffering my CF can induce with respect to renal function, from the recurrent infections that turn my urine cloudy and foul to the shooting pains from inflammation and temporary obstructions in the ducts.

Yet I have learned to think differently about suffering itself, in terms of both the potential for a successful physical outcome and the likelihood of emotional value being gained by allowing the person who gave me life to continue giving to me in that way. Perhaps most importantly, I have learned not to look at myself as a “waste” of any donated kidney I might receive, whether my mother’s or a deceased person’s, because of the progressive nature of my disease. After all, you do not get to be the oldest living person with CF without taking a few risks along the way—foremost among them opening your mind to things you thought you would never consider. As more adults with CF continue surviving into late life with each passing year, it will be increasingly critical for nephrology practitioners not only to understand the challenges we face with our kidney health, but to communicate with us about those struggles in ways that promote empowerment. Centering our voices in that dialogue offers a hugely impactful and readily achievable specific aim in support of that broader goal.

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