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Neverland: A Critical Autoethnography of Aging with Cystic Fibrosis

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Abstract

In this autoethnography, I analyze stereotypes and misconceptions about people with cystic fibrosis (CF). I examine these illness representations and their social underpinnings through critical analysis of my journey to conclusive diagnosis with CF after first being tested for the disease in early life, and the events that have followed from that turning point. Using experiential data and prior research, I explore and refute harmful misconceptions about life with CF. I challenge the notion that people with CF never grow old. I also contest the idea that people who receive conclusive diagnoses during adulthood only then transition into patient identities. In doing so, I compare and contrast my own experiences with evidence from other patient narratives. I engage lived experiences from both outside and within the CF community to explore intersectional perceptions of aging. By giving voice to the diverse realities of aging with CF, I illuminate larger gaps in the illness management literature and the health care services it informs. I conclude that narrow representations of illness experience can ultimately impede quality health care and social support for people aging with CF, and that centering diverse patient voices can positively transform both clinical and community experiences.

Keywords

Chronic Conditions, Illness Management, Critical Autoethnography, Aging, Cystic Fibrosis

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Neverland: A Critical Autoethnography of Aging with Cystic Fibrosis

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In this autoethnography, I analyze stereotypes and misconceptions about people with cystic fibrosis (CF). I examine these illness representations and their social underpinnings through critical analysis of my journey to conclusive diagnosis with CF after first being tested for the disease in early life, and the events that have followed from that turning point. Using experiential data and prior research, I explore and refute harmful misconceptions about life with CF. I challenge the notion that people with CF never grow old. I also contest the idea that people who receive conclusive diagnoses during adulthood only then transition into patient identities. In doing so, I compare and contrast my own experiences with evidence from other patient narratives. I engage lived experiences from both outside and within the CF community to explore intersectional perceptions of aging. By giving voice to the diverse realities of aging with CF, I illuminate larger gaps in the illness management literature and the health care services it informs. I conclude that narrow representations of illness experience can ultimately impede quality health care and social support for people aging with CF, and that centering diverse patient voices can positively transform both clinical and community experiences. Keywords: Chronic Conditions, Illness Management, Critical Autoethnography, Aging, Cystic Fibrosis

Introduction

In a previous autoethnography (Nowakowski, 2016b), I outlined the duality of social constructions of chronic conditions with visible consequences. I used an illness as deviance framework to analyze experiences of visibility and invisibility within different social environments and contexts and illustrated connections between these two components of the visible chronic illness experience. Within this framework, I explored how representations of chronic conditions are formed and adapted, and how these representations can both offer valuable information about what people with specific diseases experience and reproduce structural inequalities that deepen the disadvantage associated with chronicity.

When I wrote the original article, I incorporated unique insights from my life with a chronic condition that was both severe in symptoms and ambiguous in diagnosis. However, I also mentioned that my disease was very similar to cystic fibrosis (CF), a genetically inherited disease of the mucous membranes that causes severe damage to multiple organ systems. Indeed, the question of whether I had CF had been raised at multiple times. I was even cared for in childhood by a doctor who was convinced that I had the disease but received no follow up after early adolescence. About three months after my article appeared in print—just before my 33rd birthday—I was in fact diagnosed with CF. Several weeks later, I attended an annual conference to connect with other scholars doing qualitative health inquiry. There I presented my research from that first article to a diverse group of researchers who championed the value of autoethnography for critical analysis of illness experience. Critical autoethnography lends

unique perspective to analysis of lived experience by both amplifying and challenging the author's personal standpoints (Holman Jones, 2016). Rather than presenting the article in its original context, I chose to contextualize and reflect upon it through the lens of the new information I had learned about my disease.

Creating dialogue around the simultaneously unsurprising and shocking reality of being diagnosed with CF at 33 years old allowed me to enrich the concept of duality that played such a key role in the original manuscript in three different ways. First, it elucidated how receiving a more conclusive diagnosis often assuages some ambiguities of the illness experience while enhancing others and introducing new ones. Second, it illuminated the fact that many people do not realize it is possible to age substantially with CF, let alone meet anyone who can help to debunk the notion that people with this condition never get old. Third, it exposed how other people living with CF may simultaneously connect with other patients on a very fundamental level and know very little about their individual experiences—of aging or anything else. All of these additional axes of duality reflect back on the central notions of visibility and invisibility that anchored the original article.

I realized in that moment that the unlikely sequence of events leading to my eventual diagnosis with CF represented a unique and valuable opportunity to enrich the qualitative literature on chronic illness further. I was living with something quite literally construed as a never event: surviving to age 33 with a relatively mainstream physical presentation of a disease that often kills in childhood, and then finally getting a diagnosis after more than three decades of false starts. My CF had been lurking inside of me that entire time, simultaneously visible and invisible to both myself and others. Likewise, the permanent damage done to my body by those 33 years is sometimes visible to others and sometimes not. But what struck me most of all was realizing, as I followed this thread of inquiry ever deeper, that I had become an invisible population within a very visible one, largely because of inaccurate stereotypes of how CF presents in the body and impacts people's lives.

By the time I received my conclusive CF diagnosis and learned about my underlying genetics, I had been working as a medical sociologist and public health program evaluator for nearly a decade. I had held many staff positions at two different universities, earned a PhD in the process, transitioned into a specialized faculty position, and then gotten promoted into a ranked faculty position. I had at no point in my adult life worked less than full time, and I had spent much of my unpaid time on service to others with chronic conditions. In the process of developing my professional life in these ways, I had encountered many people who asked if I had CF. I had sometimes also fielded questions about stereotypes and inaccuracies in perceptions of the CF from people who assumed that I had it. When I did receive a conclusive diagnosis of CF just before my 33rd birthday, I found myself inundated with the misconceptions and fears of many people in my life. I noticed a pattern of people assuming that being diagnosed with CF meant that my health would continue to decline rapidly. Yet my conclusive diagnosis actually allowed my health to do the exact opposite. As I began to access comprehensive, guideline-based CF care, my health began to improve markedly. Indeed, reaching old age began to seem like a more realistic prospect for me after receiving a conclusive diagnosis allowed me to receive health care that met the full scope of my needs.

In this article, I thus examine stereotypes about CF and their social underpinnings through critical autoethnography of my journey to conclusive diagnosis and the experiences that have followed since and situate these analyses within the broader context of aging with chronic illness. Critical autoethnography offers unique insight into the intersecting social and cultural contexts of life journeys, including illness management (Boylorn & Orbe, 2016). I analyze and refute the notion, commonly held even among people living with CF themselves, that the rare individuals who get diagnosed late always experience blissful unawareness that they are living with a ticking time bomb prior to that point. Rather, I engage lived experience

and sociological theory to illustrate that although aging with CF after inconclusive diagnosis and improper management in childhood may be a “Neverland” of sorts due to the rarity of this event, neither the few of us walking that specific path nor the general population of people growing older with this disease have much in common with Peter Pan. I engage autoethnography as a tool for giving visibility and voice to the realities of aging with CF. In the process, I illuminate larger gaps in the illness management literature by exposing both between-group and within-group dynamics of disease representation that can ultimately harm patients by closing the minds of those who seek to provide clinical care and social support.

Background

CF is a genetically inherited disease that causes progressive and often catastrophic damage to multiple organ systems as people age (CF Foundation; <http://www.cff.org>). In essentials, CF is caused by mutations on a specific gene that are associated with the pathology of the disease (Zielenski et al., 1991). Life scientists have presently identified more than 1,000 variants on the cystic fibrosis transmembrane conductance (CFTR) gene that are associated with the disease (National Institutes of Health, 2008). All of these mutations cause changes in the functioning of chloride channels, which help move water into and out of cells in various tissues, resulting in the production of sticky mucus (National Institutes of Health, 2008). More than 30 common CFTR gene variants are commonly screened for in genetic testing for CF (Bobadilla, Macek, Fine, & Farrell, 2002).

However, no single variant or combination of variants is either necessary or sufficient to cause the disease (Knight, 1991). Early research on CF suggested that only people with two identical copies of the same variant could present with active symptoms (Davis, 2006). Yet more recent inquiry and a preponderance of clinical evidence have revealed that both people with only one copy of a single variant and those with multiple variants on one side of their genomic tree can develop both the underlying disease and its common consequences (Johansen et al., 1991). In fact, some studies now suggest that people who have no recognized CFTR mutations whatsoever can still present with many of the core symptoms of CF (Groman et al., 2002).

The core feature of CF is production of mucus with a texture similar to rubber cement (Koch & Høiby, 1993). Over time, this sticky mucus harbors harmful bacteria and occludes ducts within multiple organs (Tsui & Durie, 1997). Tissues become scarred and eroded by persistent infections and blockages to the point that they ultimately stop functioning (Tizzano & Buchwald, 1995). This process results in breathing problems, nutritional challenges, urinary and bowel issues, endocrine disruption, physical disability, and chronic pain (Emerson et al., 2002). It can also result in damage to systems not directly involving mucous membranes, such as the bones and connective tissues (Aris et al., 1998). Organ failure is a common cause of death among people with CF (Emerson et al., 2002). Complications like organ failure can result from both the underlying disease mechanisms and the autoimmune problems they cause (Høiby & Schiøtz, 1982). Although advancements in health care over the last several decades have increased life expectancy tremendously, CF patients in the United States still have a median life expectancy of 47 years as of March 2018 (CF Foundation; <http://www.cff.org>).

Most people with CF are diagnosed very early in life, usually with a sweat conductivity test (Stern, 1997). Late diagnosis of CF is comparatively rare, with only a handful of people each year getting diagnosed after age 18 in the US (Rodman et al., 2005). Delayed diagnosis may be partly a selection effect, as remaining undiagnosed limits opportunities to access evidence-based care. The ability of the body to adapt sufficiently to survive long-term with undiagnosed CF may hinge in part on the specific gene variants a person has. However, both cases of people remaining relatively asymptomatic with two copies of one or more variants *and*

cases of people experiencing severe symptoms with only one copy of one variant have been observed by clinicians (Lommatzsch & Aris, 2009). In short, scholars and providers alike still have much to learn about CF and how it impacts individual patients.

Autoethnography thus offers a uniquely powerful tool for inquiry on CF because it involves the critical study of the self and one's own experiences (Ellis, Adams, & Bochner, 2011). It has been recognized for nearly 30 years as a valid and valuable method of empirical research in the social sciences (see Berger, 1990; Riley, 1988) because of the unique information and perspectives it can contribute (Ellis et al., 2011). Using autoethnography for empirical inquiry can greatly enrich bodies of research that otherwise rely on other methods (Anderson, 2006). Championing and conducting autoethnography can also help to prevent bias by forcing scholars to consider and deconstruct their own standpoints both for social science generally (Ellis et al., 2011) and for embodied health research specifically (Murphy, 1987). Building on early autoethnographies exploring acute illness, scholars increasingly engage autoethnography to further understanding of chronic disease (Karnilowicz, 2011).

Because the burden and experience of chronic disease often differs sharply by socioeconomic status and related characteristics, health scholars have a responsibility to use a variety of tools to amplify the voices of marginalized populations, especially those whose circumstances they understand firsthand (Biber & Leavy, 2007). People with CF experience marginalization in clinical care because their condition remains an "orphan disease" with few effective therapies (US Food and Drug Administration, 2018). The perception that people with CF do not grow old intensifies this marginalization. By contextualizing my life with CF through my experiences as a female, agender, multiethnic, and disabled person, I was able to offer examples of complex marginalization that can directly inform person-centered care. Likewise, I explored my privileged positions of whiteness, affluence, and education to present intersectional insights on inequity in the CF community. Painting an impactful portrait of illness experience requires understanding these nuanced social and cultural systems of advantage and disadvantage (Conrad & Barker, 2010).

Autoethnography has also provided a useful tool for many scholars seeking to improve the evidence base about visibility dynamics in social responses to chronic illness. Such approaches to autoethnographic inquiry frequently engage the notion of stigma, which comprises adverse value judgments and subsequent action on these conclusions (Goffman, 1963). Illnesses that involve both visible and invisible elements present particularly exciting opportunities for autoethnographic inquiry because they can illustrate multiple types of discrediting and resultant stigma (Wilbers, 2015). Likewise, autoethnography can illustrate how people with chronic conditions balance self-management behaviors with intersecting social roles—a process that often creates conflicts between multiple identities held by patients simultaneously (Strunin & Boden, 2004). Reflecting on the intertwining perspectives of life with and scholarship on CF proves particularly instructive in this regard, all the more so because CF often presents with a combination of visible and invisible symptoms and thus creates complex social and behavioral expectations.

Outside of research settings, adults with CF have long been exploring their experiences in writing and sharing them with others. As the median age of survival has increased for people with CF within and outside the United States, more of these stories have begun to contain specific information about what it is like to grow older with the disease. Books like Jay Geronimi's *Can't Eat, Can't Breathe...*; Laurel Rothenberger's *Breathing for a Living*; and Andy Lipman's forthcoming *CF Warriors* all engage the concept of aging and situate it within the context of living with progressive disease. These layperson accounts may be considered precursors to formal autoethnographic inquiry (see Julia, 2003) on the process of growing older with CF, which at present has never before appeared in the research literature on aging with chronic illness.

Methods

Type of Inquiry

I used critical autoethnography to both reflect on a variety of lived experiences related to CF and late diagnosis thereof. Critical autoethnography offered an ideal framework for conducting inquiry on my experiences of aging with CF before, during, and after conclusive diagnosis. Using critical autoethnography proved ideal for two principal reasons. First, it required me to both robustly engage and carefully challenge my own personal standpoints as a patient and scholar (see Holman Jones, 2016). Second, it required me to situate my own experiences within broader social and cultural contexts (see Boylorn & Orbe, 2016).

I followed both general recommendations for using qualitative methods to discover important dynamics in illness management (Charmaz, 2000) and specific recommendations for using personal biography as a tool for inquiry (Bury, 1991). As in my earlier work (Nowakowski, 2016a), I once again used a narrative approach. This inquiry process allowed me to synthesize my perspectives as researcher and case example, and accommodated variability in potential interpretations of the specific experiences I analyzed. My overall goal was to stimulate interest and curiosity concerning the diverse possible experiences of people with late diagnosed CF and poorly understood chronic conditions in general, not to paint a deterministic portrait of what other individuals in these groups may experience at a given time.

Participant Engagement

This study had one direct participant (myself as both scholar and subject) and multiple indirect participants (people who interacted with me in some way concerning my health). Although other people did thus contribute some data, the method of inquiry here was exclusively retrospective. I used critical reflection on lived experiences that had already happened by the time I began taking notes for this manuscript. As a result, this study did not require IRB review because it was considered oral history (see Shea, 2000). Standards for confidentiality in reporting oral history results are both important and unique. I protected participant confidentiality in this manuscript by never identifying any of the individuals described in the manuscript by name except for myself. I also created no supplemental files linking specific clinicians or community members to the study. To promote intersectional inquiry, I did try to provide some informed conjecture about contextual factors that might be salient to particular individuals who interacted with me. However, I did not provide a sufficient level of detail about any other person's life to allow for identification of that specific person by readers, except in one specific case where a close friend granted me permission to discuss our social relationship and his experiences with CF.

Data Collection

Data for this study were collected entirely through retrospective inquiry via an oral history approach. Oral history constitutes an evidence-based and validated method for autoethnographic research (Ellis et al., 2011). It can capture the diversity and complexity of individual illness experiences with a high level of detail. I used methods successfully employed by other medical autoethnographers to compile and analyze my data, bridging these general approaches with the foundational content generated in my prior work. As with those prior oral history based publications, critical reflection on past events formed the basis of my approach (see Rier, 2000). This time, I used the final draft of prior articles as a sort of field journal,

annotating each portion of the content with new insights gained from obtaining and accepting a CF diagnosis.

Because this study originated in a desire to build directly on my previous work, I began with a more defined set of guiding threads for inquiry. These questions are detailed below. To prepare for writing the manuscript, I also returned to the literature on autoethnographic research and the use thereof for health-related topics. I focused on research amplifying patient stories as well as gaps in those narratives (see Charmaz, 2002). In addition, I began exploring both the research literature on CF diagnosis and management and the patient literature on lived experiences of that specific disease. These explorations enabled me to craft two questions for exploration through critical reflection and engagement of relevant theory. Specifically, I wanted to reflect upon:

- (1) What misconceptions about CF limit visibility and understanding of patient experiences among people who do not have the disease?
- (2) To what extent do these dynamics differ, if at all, among people who *do* have CF themselves?

In interpreting my results for these two threads of inquiry, I focused on how clinical and community discourse on CF could change to better affirm the needs of individual patients in light of these findings.

Data Analysis

After generating a new set of field notes by marking up final versions of prior articles, I used a grounded theory approach to analyze these notes. Specifically, I performed content analysis with open coding, grouping distinct observations into shared categories organized by major themes (Charmaz, 2006). In the process, I took additional notes on theoretical traditions and pieces of research literature that would lend context to my own observations. I organized my results section using both the guiding questions I began with and the thematic categories I outlined during the content analysis process, then used my notes on relevant theoretical and empirical literature for discussion of findings for each thematic topic within each overarching question.

I tried to create the highest level of rigor possible in interpreting my field notes. Using previously published autoethnographies of chronic disease as well as methods literature on autoethnography itself proved extremely helpful. Yet likewise, I focused on the unique contributions that each new autoethnography can make (Murphy, 1987) even in cases where that meant diverging sharply from evidence and interpretations presented in prior publications. As in all of my autoethnographic work, I reflected frequently and explicitly on the value of my interwoven perspectives as researcher and patient (Liggins, Kearns, & Adams, 2013). I continued my practice of requesting input from peers and mentors with rich experience in qualitative inquiry and critical autoethnography. This time, I also solicited guidance and feedback from other people living with CF, some of whom were also scientists or public health professionals in their own right and some of whom were not. This last part of the inquiry process proved especially helpful in identifying limitations of and blind spots in my own insights.

Presentation of Results

I organized my findings first by guiding question and then by thematic area within each question. To contextualize each portion of the back matter, I returned to the core theoretical

traditions I drew on in the original manuscript. These included structural functionalism (Parsons, 1951); the associated concept of illness as deviance (Twaddle, 1981); and the related theories of how illness representations contribute to stigma (Goffman, 1963). I also incorporated a variety of new literature, drawing in particular on writings related to CF specifically. I engaged research literature from scholars in the interdisciplinary sociomedical sciences, as well as written and filmed accounts from other people living with CF. These theoretical and empirical source materials provided context for my own case examples.

Whenever possible, I took an intersectional approach to contextualizing my own data and findings. Reporting the perspectives of a single person inherently poses problems for generalizability. Although I sought to achieve a higher level of population context by incorporating information from accounts of other people living with CF, I did not set out to comment on specific things that any one person with the disease does or does not experience. Rather, I focused intensively on generating valuable insights for inquiry from my own observations, and on articulating how these critical perspectives could be used to enhance clinical care and social support for other people with CF whose experiences may both overlap with and diverge from my own. In both clinical and social settings, affirmation requires seeing and embracing people as they are in the moment (Thoits, 1995).

I thus strive to make my own writing reflect the experiences of others accurately. For example, I use gender neutral pronouns in referring to people whose pronoun preferences I do not know. In this specific manuscript, I did not need to use neutral pronouns for any particular individual except myself but continue to advocate for the general practice of defaulting to neutral language in cases of unknown preferences. I also err on the side of caution in reporting attributes such as race and sex, describing my perceptions of people rather than trying to guess at how they identify. This practice has become commonplace in critical autoethnography (Ellis et al., 2011) for good reason. Specifically, it helps to amplify the voices of other people and preserve the agency of individuals experiencing marginalization (Biber & Leavy, 2007). Because CF frequently causes disability and associated perceptions of people with the disease as lacking in power and agency, using open and person-centered language was essential in reporting results.

Findings

Guiding Question 1: What misconceptions about CF limit visibility and understanding of patient experiences among people who do not have the disease?

You can only have CF if you have two or more genetic mutations.

When the genetic underpinnings of my particular presentation of CF were identified, I found out that I was literally the only documented case of the disease in the United States with that specific genetic mutation (c.1584+36A<G). I had none of the most common CF mutations, although years prior, the screening I did with 23andMe had indicated nonspecific problems with the coding of my CFTR genes. I also only had that one single mutation, a substitution at location 1584.

Basic science on CF is just now evolving to recognize that certain types of mutations can effectively turn off a properly coded CFTR gene on the other side of a person's genomic tree. Most of these variants are likely something called intron mutations that interfere with the splicing of genes themselves (Gargouri, Lazowska, & Slonimski, 1983) or messenger RNA, the small molecules that communicate between a person's base genetic material and cells in the early stages of protein production (Reed & Maniatis, 1985). However, intron mutations are rare. Finding out my genotype thus marked me as an outlier in the data. Theoretically, I should

have been an asymptomatic “carrier” rather than having a very mainstream CF phenotype (Kulczycki, Kostuch, & Bellanti, 2003). It also made visible the specific issue that caused me to have symptoms in the first place, lending validity and coherence to my experiences and the complexity of my journey.

One of the insights I gained concerned the precise nature of my genetic mutation. At location 1584 on the CFTR gene, any aberration is likely to produce difficulties with the “residual function” of the protein the gene helps to produce (Welsh & Smith, 1993). If this mutation impacts the functioning of both CFTR genes, it can produce the full disease phenotype but also foster a slower overall disease progression than a more classic genotype would (Green et al., 2010). Understanding some of the subtleties of how intron mutations on the CFTR gene may operate helped me to make theoretical sense of why I had survived multiple complications from CF, and specifically of why my lung function had rebounded once I was put on inhaled corticosteroids. However, it could not fully explain my individual experience with the disease—no single factor can.

Your genes determine your fate.

When the CFTR gene was discovered in 1989 (Riordan et al., 1989) scientists quickly identified a particular deletion at location 508 that was thought to be responsible for all cases of CF disease (Zielenski et al., 1991). Several years later, it became apparent that the 508F deletion could not begin to account for everyone with the CF phenotype—or for survival outcomes in people who had that mutation versus those who did not (Riordan, 1993).

As I was beginning to integrate within the CF community, I had the good fortune to meet a couple of fellow health research professionals living with the disease. Both had two copies of 508F. One was a public health scholar the same age as me and had no history of transplantation or even signs of incipient organ failure. The other was a long-term care scholar and administrator almost 30 years older than me and had undergone a double lung transplant nearly two decades prior. Both were in good health and enjoying a life that included plenty of physical activity. I found it uplifting to meet people with the double delta 508F genotype who were aging in good health, whether with the aid of a transplant or not. I also found it depressing that with a rare mutation that should theoretically have much milder impacts, I wound up worse off in many ways—damaged kidneys, heart, vascular system, etc. I still struggle with this cognitive dissonance (see Festinger, 1957) today.

Likewise, I heard some stories from my care team about older people with CF that surprised me. Perhaps the most astonishing of these accounts was the tale my CF doctor told me about a patient he cared for during residency. This individual was in her late 60s and had been very active all her life, with no major physical health challenges. Recently, however, she had been feeling short of breath while swimming. After ruling out several other possible issues, my doctor suggested doing a blood test to see if by chance she had any CFTR mutations. It turned out that she had two copies of the 508F deletion, which at the time made little sense to clinicians given her complete lack of symptoms until late midlife.

Scientists have begun to explore long term nonprogression of CF in people with the double delta 508F genotype (Collawn, Lazrak, Bebok, & Matalon, 2012). A cohort of five people with this genetic profile at a large CF clinic in Australia is being followed prospectively because the doctors caring for these patients noticed they showed no signs of disease progression over long periods of time. Their clinicians partnered with geneticists to explore whether other things in these patients’ genomes could help to explain why people with such a classic CF genotype experienced little worsening of their symptoms over time. This research team discovered that certain other types of mutations on other genes that impact the

transmission of sodium across cell membranes can protect against some of the worse effects of CF on the body (Agrawal et al., 2017).

I would later share a manuscript describing findings from that research with a graduate level genetic counseling class I spoke with at a university. Some of the students had interest in CF genetics, and the professor requested that I come and share my story as an introduction to a research-based discussion of how the science is changing. Students asked many probing questions that day about CF genetics and aging, a key frontier in the evolving scientific literature on the disease. I felt both enthused about the questions they were asking and somewhat helpless in answering them. I do not know how other people with my mutation age, as a population, because I am the only documented case of the disease in the United States associated with this genotype.

After I gave that talk, I reflected on it with a friend from the general CF Facebook group—a young bioscientist currently finishing her undergraduate work. She has one copy of 508F and one rare mutation and has enjoyed high lung function her whole life as well as relatively mild impacts to other systems. Although her adherence to treatments has always been superb (see Sawicki, Sellers, & Robinson, 2009) she said she also wonders if she may have some of those beneficial mutations on other genes. This exchange made me reflect on how the process of aging with CF often includes keeping up with current scientific developments and thinking about what new knowledge may mean for our own care. It also reminded me that there is no way to predict a person's lung function at a given age, or over time, based on genotype alone.

Adults with CF cannot have high lung function.

The biographies of people aging with CF lend ample insight into the diversity of how and to what extent the disease impacts the lungs. My own case is fairly typical in the sense that without medication or other management techniques, my symptoms fall about 50 percent into the “lung” area and about 50 percent into the “other” area, with the latter consisting mostly of gastrointestinal and pancreatic function issues. With medication, however, my worst daily challenges tend to come from my poor digestive and exocrine function. I spend a lot more time running back and forth to the bathroom than I do coughing on an average day.

As I have become more integrated into the CF community, both in a general social sense and through specific advocacy and education efforts, I have met numerous other patients of similar age with high spirometry (a way of measuring how much air the lungs can expel at once, see Gustafsson, Aurora, & Lindblad, 2003) numbers. Some of my friends who can still blow a triple-digit number on their forced expiratory volume assessment have relatively few lung symptoms day to day, and have oxygen saturation levels indistinguishable from those of people without CF. By contrast, I blew a full 100 percent on my most recent spirometry—a higher number than I had ever seen in my adult life prior to that day—but routinely experience oxygen saturation levels several percentage points below “normal.” My levels are high enough that I do not need supplemental oxygen, but low enough that I clinically develop hypoxia if at elevation or experiencing additional inflammation in my lungs from a viral infection.

This immense diversity in lung function outcomes helps to explain why in the CF community today, we see stories about both children dying before adolescence and adults thriving into old age. It also gives context to the confusion that people unfamiliar with CF often show about who needs oxygen supplementation and who does not. When I gave my presentation on my prior research, I did a quick straw poll in the room about perceptions of surprise at me not using oxygen despite being 33 and living with CF. Almost every person in the room raised their hand. Yet most of the adults with CF I know—whether younger than me,

the same age, or much older—do not use supplemental oxygen 24/7, and many do not use it at all into midlife and beyond.

People with CF do not grow old.

The dual visibility and invisibility of my lived experiences of living and growing older with my disease became all the more true as I began to meet other people with CF whose condition was not diagnosed conclusively until adulthood, many of whom had few symptoms until middle age. By contrast, I had been ill since the earliest years of life and had nearly died from complications of the disease several times without ever knowing for sure that I had it. My adjustment to the sick role (see Nuttbrock, 1986) began very early in life despite my lack of conclusive information about why I was ill. I thus experienced a lot of cognitive dissonance in adjusting to the fact that I was such a rare story—a “unicorn,” as my CF doctor once called me—even after being given the correct diagnosis.

People both within and outside of the medical community were stunned that I had survived for so long without coordinated CF care, and that I was able to regain some of the lung function I had lost. Over time, I would also stabilize with my kidney and vascular symptoms after beginning to use some different medications and modifying my diet. My team of clinicians where I had been living since receiving a conclusive diagnosis, helped me make sense of some of the physiological factors that worked in my favor. I am still exploring the unique attributes of my genotype as a research participant with our partner clinic. Psychologically, however, I required quite a bit of time for adjustment.

When I first began working with my CF doctor and extended care team, we spent a lot of time discussing the fact that my odds for long term survival were likely to improve now that I was getting better care. My CF doctor openly expressed empathy for my feelings of anger about the past, disappointment about the present, and uncertainty about the future. Within the space of a few weeks I was diagnosed with digestive enzyme deficiency, thickened airways, diseased kidneys, lung nodules, sinus polyps, and failing blood vessels. I already knew I had a heart condition, circulatory problems, nerve damage, and extensive scarring inside many of my organs.

However, as we were gathering all this diagnostic information my doctor and I also worked on adjusting my medications and other management strategies to meet my needs as an adult with CF. For example, I started to see positive changes after being placed on pancreatic enzymes and special vitamins for CF patients. With more aggressive physical therapy, my lung function increased to the point where I blew that 100 percent number on a recent spirometry test. My weight remained low and still does today even with a higher dose of enzymes, but I feel exponentially stronger than I did before. My sleep has become more restful and I do not spend every day feeling as if I am about to lose consciousness. I have more good days than bad ones lately (see Charmaz, 1991).

At the same time, I am still living with a disease that will eventually kill me. I am now doing so while adjusting to the idea that the disease may take so much time to kill me that my lifespan will be about the same as it would have been without CF. My doctor’s admirably aggressive posture on this is that I should not have my life expectancy shortened by even one day because of my disease. He reminds me that our focus should be on making sure I can get every single one of those days, and to be able to enjoy as many of them as possible with a high quality of life. This has been amazing to hear and absorb, and also heartbreaking because of how it makes me reflect on all the times I almost lost my life as a younger adult (see Rier, 2000). It still feels particularly painful to think back on the years I spent in a body spiraling toward death and wondering when I would get my last second chance to live.

I do not know if the trauma of those years will ever fully fade. However, I do know that it will always be an integral part of my own understanding of what it means to age with CF. As I became more integrated and active within the CF community, I would quickly learn that people diagnosed much earlier in life also experience many of the same sources of dissonance between having good days in the present and fearing what may happen in the future (see Asbring, 2001). By contrast, people with CF who were diagnosed early may or may not experience the same feelings of anger and hopelessness about having “lost time” in our care histories (see Baumgartner, 2007).

Guiding Question 2: To what extent do these dynamics differ, if at all, among people who do have CF themselves?

People with CF usually understand that presentation of the disease varies.

The rise of the Internet has greatly facilitated social connections between people with CF, which previously proved difficult because of the “six foot rule” imposed on patients (Lee et al., 2017). More of a guideline than a formal rule, this term refers to the recommendation that people with CF never come within six feet of one another even if wearing masks (Saiman, 2011). The six foot rule helps to protect people with CF from colonization by one another’s lung bacteria. In the interim time between the discovery of potential harms from contact between patients and the rise of the Internet, the six foot rule also caused many people with CF to feel isolated and unsupported (Johnson, Ravert, & Everton, 2001). Indeed, a clear theme about aging with CF that appeared in both my own introspection and my dialogues with others in the community is the importance of spending time in spaces where we feel “normal” (Josefsson, 2005).

A lot of attention gets devoted in online CF communities to discussion of what “normal” even means for people living and aging with the disease. We tend to talk about our symptoms as a way of introducing ourselves to others, and frequently use language like “is this normal” or “am I the only one” to initiate discussions of our experiences. Sharing our mutations and comparing them with those of others is a common “icebreaker” activity in online CF communities. I remember feeling very excited about learning my own genotype because amongst other things, it enabled me to participate substantively in these discussions. Yet learning more about my genetics also showed me something valuable about the diversity of how the disease presents and progresses as people age. I have met a few other patients online who have residual function mutations at or around location 1584 on the CFTR gene. However, none have symptoms or stories that exactly mirror my own.

The online CF community appears to do a very good job of operationalizing symptom clusters as axes on which specific patients may load more or less strongly. Understanding that people can have terrible gastrointestinal symptoms and mild lung symptoms—or the reverse—is quite common and consistent. Knowledge about secondary complications from the disease, such as kidney dysfunction and vascular problems, is likewise present among participants in patient online communities. However, my own explorations suggest this knowledge is much less consistent from person to person. A lot of people with limited clinical education post questions about whether something they are experiencing could be related to their CF. This is often a difficult question to answer even for people with extensive training on CF management, as the disease impacts a variety of different systems in the body (Zielenski, 2000). Yet a lack of general knowledge on what types of complications fall within the scope of CF itself—for example, confusing Type II diabetes with CF related diabetes—appears to be somewhat widespread.

Another major development concerning disease representations and aging that accelerated even during the year-long period I spent doing this autoethnography is racial diversity in visualizations of people growing older with CF. I had begun seeing pictures of racially diverse younger patients in the publicity materials for CF care centers, but until recently saw few such representations of patients well into their adult years. Projects like Ian Pettigrew's "Salty Girls" photo series (2012), as well as the photo sharing features nested in many social media platforms that host patient online communities, are helping to change these narrow perceptions of what a person aging with CF can look like.

The differences in bone structure, skin tone, and other physical characteristics that get socially coded as "race" can also confound our sense of what is physiologically "normal" for people who are growing older with CF. For example, a discussion occurred on one of the Facebook groups I participate in about pseudoclubbing, a phenomenon where someone simply has thicker fingertips and curved nail beds from birth. This occurs in people of all races but is more common among Black populations (Callemeyn, Van Haecke, Peetermans, & Blockmans, 2016). Likewise, not all people with CF ever experience "true" clubbing (inflammation in the fingertips that arises in response to low blood oxygen levels increasing production of a hormone called prostaglandin E2) even if they survive into late life (Lemen et al., 1978). So, the concept of what a body with CF can look like as it ages may vary intersectionally across multiple characteristics, including race.

Body mass and shape are other factors getting increased attention in the CF community right now with respect to aging. A frequent topic of discussion—and tension—within online CF groups is the fact that not all people with CF have thin bodies. The stereotype of adults with the disease as being very slender has roots in the prevalence of exocrine pancreatic insufficiency among people with CF. This condition involves the pancreas not releasing enzymes into the intestines to help people digest food (Domínguez-Muñoz, 2011). In CF, this is caused by blockages in the small ducts inside the pancreas (Colombo et al., 2009). Developing diabetes can also cause weight loss before treatment begins (Lanng, Thorsteinsson, Nerup, & Koch, 1992). CF related diabetes develops when the body cannot move the insulin it produces into the bloodstream, again because of blocked ducts (Mackie, Thornton, & Edenborough, 2003). However, not all adults with CF have either of these conditions. The impact of both conditions on body size and shape also occurs relative to people's individual baseline. A person who would otherwise have been very heavy might be thinner if they develop CF related diabetes or pancreatic insufficiency, but they may never be thin. Some CF patients with larger bodies have experienced skepticism and outright discrimination about the validity of their diagnoses because of their weight—a terribly dangerous proposition for aging with the disease, as time often translates to health in managing it.

These are specific examples of the broader discourse concerning bodily diversity in the CF community, and how that translates to popular consciousness of what people with the disease look like as we move through life. Assessing these anecdotes and thinking about their broader implications lends insight into potential sources of cumulative inequality (see Ferraro & Shippee, 2009) in aging with CF—and how to address them. Meeting physically diverse people aging with CF has enabled people both within and beyond the community to broaden their concept of what older people with the disease look like beyond the stereotype of a frail white person with clubbed fingers, transplant scars, and an oxygen tank.

Many people with CF know that some patients survive into old age.

Getting opportunities to meet middle aged and older people with CF has become much easier over time, both because of improved survival and diversified technology (Lee et al., 2017). Patient online communities—whether hosted on dedicated sites such as PatientsLikeMe

or nested within social media platforms such as Facebook—have opened up myriad opportunities for adults with CF to connect and form friendships. Advocacy and education activities may be interspersed with these social exchanges. This has been the case for me, lending an additional layer of closeness and meaning to the relationships I have formed. I have also been surprised by the extent to which these “spaces of normalcy” online have facilitated the creation and sharing of creative content related to aging with CF.

My inquiry into misconceptions about growing older with CF naturally led me to explore creative representations of adults with the disease—both those I found on my own and those I accessed after fellow members of patient online communities shared them within those groups. In addition to multiple books on living well into adulthood with CF, I found other forms of creative engagement addressing the biographies of aging adults with the disease. Two that stood out were the aforementioned “Salty Girls” photo series and the work of mixed media artist “Blinded for Review.” The latter is the creator of “Google 65 Roses” and other installations about CF and the experience of physical change induced by the disease.

I wound up becoming close friends with “Blinded for Review” after we met on a general Facebook group for adults with CF, and bonded over our shared interest in visual art. As not only an artist himself but also an art educator with a background in the social sciences, he was able to share with me about his journey towards understanding his aging body through art in language that resonated with my own training in medical sociology and social psychology. A key theme in his recent works has been exploring and navigating the experience of embodiment following lung transplantation, which he successfully underwent some three years prior to our meeting. He explained his feelings on aging and his sense of his body on that timeline in a way that resonated with my own descriptions of being “twenty-three plus four” (Nowakowski, 2017) in a previous book chapter about my journey with chronic illness. I used that language to describe my sense of my life bifurcating into the time periods before and after my admission to intensive care.

Likewise, he had a sense of rediscovering his body and what it could do as he continued to grow older that resonated strongly with me—all the more so because we were only a year apart in age.

Yet my friendship also showed me how different our experiences had been, even though we had both wound up with permanent damage to our bodies as most people with CF do. He had been diagnosed conclusively early in childhood and was revealed to have a very common genotype for CF once he underwent genetic testing as an adult. He had experienced many similar physical challenges to those I faced myself, but with none of the ambiguity about why he was sick or what could be done to help him. My particular experience with late diagnosis afforded me none of the sense of coherence that people with early conclusive diagnoses get (Jutel, 2009) and thus introduced some specific mental health challenges despite my physical experiences often being very typical.

Late diagnosis is still highly contested and poorly understood within the CF community.

Getting more integrated into the CF community and forming deep friendships with other patients both affirmed me as a “normal” person and made me feel isolated by my specific trajectory with the disease. I finally met another person with a story like mine while facilitating my first-ever miniconference breakout session for the CF Foundation. One of the participants in my group happened to be a fellow person who had been sick for her entire life but not diagnosed properly until after age 30. Our stories were almost eerily similar, all the more so because we shared that we had both experienced intimate partner abuse and that she lived within a mile of the house in which I grew up. The subject of domestic abuse and CF could easily populate multiple research manuscripts; one thing I have learned with harrowing

consistency is that I am hardly alone in my history of being treated like a lesser person by a previous partner for having an incurable chronic disease.

Relative to this specific exploration, what I learned from meeting this conference breakout participant was that some people with a later diagnosis are reluctant to share that detail because they fear being labeled as attention-seeking or overdramatizing their illness. Both this individual and myself had extensive histories of poor outcomes, including hospitalizations in intensive care and permanent damage to organs other than our lungs. She was older than I was—in her early 50s—and just beginning to navigate dating and relationships again after leaving an abusive relationship. Both of us expressed that we found it cathartic to share our apprehensions about how people would respond to and value our bodies as CF patients with a history of sexual abuse. As someone in an open marriage, I was still wrestling with those concerns on a daily basis (see Cole, 1975) despite feeling extremely secure in my relationship with my spouse.

I also realized in a very clear way how much brighter my future looks at present than it did before I got a conclusive diagnosis. Finding out for certain that I had CF opened up many anxieties and fears, both related to and separate from my history of relationship trauma. One of the biggest challenges I faced was the gradual revelation that my physical presentation of the disease is quite mainstream—I would not be considered a “mild” case despite my improvements in lung function. However, this process of trying to figure out where I even fell within the broader data stream on aging with CF proved instructive as well as frightening. I learned from others in online CF communities that this is a process nearly all adult patients go through as we continue to grow older with the disease. We continually redefine our concept of what is “normal”—a sort of illness management (Charmaz, 2000) writ large at the population level. “Classic” CF has always been a somewhat murky concept (Knowles & Durie, 2002). Even people who fall firmly into that category within the general consciousness of patient communities may fare far better over time than certain others with less mainstream presentations (Boyle, 2003).

Indeed, many of us are also becoming conscious that timing of diagnosis may mean little about either disease severity or likely outcomes from aging. My very first friend with CF, someone I had known through a colleague before I got my own conclusive diagnosis, has a relatively common set of genetic mutations and was diagnosed at a very young age. However, his impacts were milder in childhood—he was diagnosed via chloride sweat testing, but this was only done because his sister had a more severe presentation of CF. He later became eligible for a new drug that made him nearly asymptomatic. Although we are similar in age, in nearly four decades of living with the disease my friend has never once disclosed his CF to someone outside of loved ones and myself. He had never joined an online CF community or otherwise sought contact with other patients until I received my diagnosis. Even then, he felt hesitant about reaching out. His life as a “stealth” CF patient (see Lowton, 2004) fascinated me, not least of all because it challenged stereotypes about early and late diagnosis.

Adults with CF are still learning the implications of late diagnosis for aging.

Because long term survival with CF has historically been poor and options for early diagnosis of the disease have improved so much, many adults with the disease have never met a late diagnosed person who had substantial health problems during childhood. Once I joined the online CF community myself and began to share my story, I quickly became known as something of an authority on the “cautionary tales” associated with late diagnosis of phenotypically mainstream disease presentations.

I also realized that there is a lot of insistent terminology (arguing semantics over the precise wording of a concept, something that has not yet been explored explicitly in

sociological literature) being used in the online CF community concerning late diagnosis. In many patients' eyes, I was not "late diagnosed" but rather "misdiagnosed" or "improperly diagnosed" or "badly managed" in my youth. In the patient online communities where I spend most of my time, "late diagnosed" is sometimes used in a divisive way to separate the experiences of people with lifelong illness from those without symptoms who only find out they have a CF genotype because of ancillary issues like reproductive difficulties. The term gets used as an insult of sorts against people who make intensely emotional posts about their diagnosis at a late age, only to reveal that they have been asymptomatic their entire lives. I have seen my fellow community members comment on such posts with dizzying speed, criticizing the authors for engaging in self-pity when their experiences have been so much tamer than those of others.

As someone whose lack of a conclusive diagnosis in childhood cost me a great deal, I cannot say that I disagree with these critiques. Yet I also realize, as a medical sociologist, how differently people experience new information about their health if they have been sick all their lives versus if they are new to chronic illness. Doing this autoethnography has shown me that the process of adjustment—either to a new diagnosis or to novel experiences from an older one—is one of the cornerstones of aging with CF. This process of illness management (Charmaz, 2000) is hardly unique to CF itself. Yet as a progressive disease, CF requires constant reevaluation of one's health status, needs, and goals (Staab et al., 1998). It also requires constant suspicion when things appear to be going well.

Both the CF community and those on its fringes continue to learn about the implications of late diagnosis for healthy aging with the disease. Concurrently, we also continue to learn about what can go wrong as people diagnosed very early in life grow older. We talk in the CF community about "The Fear"—the idea that all it takes is one bad day or being in the wrong place at the wrong time, for a terrible downward spiral to begin. The Fear is very real and present for anyone with this disease, all the more so if we have already endured catastrophic outcomes that brought us close to death.

Getting more involved in the community showed me the many faces The Fear can take in the lives of aging patients, none more so than a friend of mine who until about two years before I met him had been the picture of health—or as close as a person with CF can get to that state. He had been employed full-time, active in competitive bodybuilding, and generally living with few symptoms as a result of excellent adherence to his treatment plan and an aggressive approach to managing complications. Then he began culturing an aggressive strain of bacteria in his lungs that required treatment with intravenous tobramycin, a strong antibiotic. The tobramycin left his lungs vulnerable to colonization by black mold. It also took the vast majority of his hearing, leaving him clinically deaf. After two scant years, a future that had at one point looked relatively uncomplicated as CF journeys had refashioned itself into something entirely different—uncharted, disorienting, and frightening.

As someone not only living with CF but also studying it, my own sense of what it may mean to continue growing older with this disease is necessarily and quite reasonably haunted by The Fear. My friend from the Facebook community who lost his hearing was much healthier than I was for most of his life. One bad day, one wrong breath, changed everything. The same could happen to me at any time, likely with an even more disastrous outcome given the substantial damage done by poor management of my CF earlier in life. I grow a little older every day with the looming awareness that one more kidney infection could take away the functioning I have left, or that breathing in the wrong place at the wrong time could steal the gains I worked so hard to make with my lung health. This is the reality of aging with CF: hope and uncertainty, both driven by copious data. Before my diagnosis was confirmed, I once referred to hope as a difficult concept for a reason (Nowakowski, 2016a). I stand by that assertion today more strongly than ever.

Discussion

Contextualization of Findings with Prior Work

Findings from this autoethnography suggest that people both within and beyond the CF community struggle to understand what it means to grow older with this disease. Even patients ourselves may be unsure of what to expect for our own physical health as we continue to age. The constant evolution of clinical resources to support our care—such as new devices and drugs—means that we must frequently reevaluate our sense of what is possible for our daily self-management. By consequence, we must also continuously assess how our mental, behavioral, and social health may change in response to both new developments with our bodies and differences in the resources we can use to manage these challenges.

Managing the social expectations of others concerning aging with CF represents a major component of the experience of growing older with this disease. This phenomenon matters not only in the interactions of people with CF with our peers who do not have the disease, but also with our fellow members of the patient community. The contested nature of late diagnosis and people's associated experiences of illness represent major barriers to accurate understanding of and expectations concerning aging with CF even among people who have lived with the disease ourselves for many years.

Preliminary informal work on experiences of aging with CF also suggests several clear pathways to building this understanding. The rise of patient online communities has facilitated diverse and close connections between people with CF. It has also illuminated the varied ways in which adults with CF make sense of their unique experiences of aging and provided an accessible means of sharing those explorations with others. Adults with CF are using a wide array of approaches to introspection about growing older, including but not limited to: journaling, creative writing, photography, painting, sculpture, musical performance, poetry, and meditation. Both the process of self-exploration itself and the ability to share that journey with others are helping people with CF to make sense of our unique biographies of aging. This includes understanding what we need from both our social relationships and our health care providers in support of continued progress toward our goals.

Strengths and Limitations of this Study

This autoethnography of growing older with CF has a number of important strengths. It amplifies the voice of someone who has aged into their mid-30s with CF and is predicted to have a long life with the disease. It showcases both the diverse and often confusing experiences of seeking and adjusting to a conclusive diagnosis with this disease, and the process of readjusting to growing older with CF. It affords insight into the duality of living with a disease that has a high fatality rate while also taking a long time to kill many of the people it affects and explores the nuances of this dichotomy through the perspectives of someone whose care became a process of making up for lost time. It also incorporates a broad and diverse knowledge of the scientific literature on CF and recent innovations in research. Drawing on my family backgrounds in biology, medicine, and genetics research as well as my own training in medical sociology, public health, and social psychology enabled me to critically evaluate my own experiences in the context of a wide variety of prior scholarship.

Likewise, this inquiry has a number of substantial limitations. Writing critically about one's own experiences always involves a certain amount of skew in perspective, as one can never step fully outside of oneself. It proves similarly challenging to write critically about one's own perceptions of others' words and thoughts, given the impossibility of knowing the full scope of experiences and contexts that underlie another person's stories. Finally, it would have

been completely unrealistic to try to capture the full range of adults growing older with CF in even just the United States, let alone the world. Here, I described my interactions with certain members of the CF community with whom I have had substantive exchanges and grown close.

Even in those circumstances, I remain conscious of my own limitations in understanding the nuances of other people's voices. This autoethnography allowed me to amplify the stories of many people who have taught me something valuable about aging with CF. Indeed, the stories I found myself contemplating and sharing constituted some of the greatest surprises I experienced while preparing this manuscript. Yet neither this nor any other approach to critical inquiry could ever enable me to speak entirely for another person. This simple truth lies at the core of why we need diverse qualitative inquiry on growing older with chronic disease in general, and on aging with CF specifically: If you have met one person on that journey, you have met one person on that journey.

Implications and Future Directions

This first autoethnography of aging with CF suggests that amplifying the voices of patients who are growing older with the disease represents both an important cornerstone of improving our collective understanding of this process, and a challenge in many other respects. Specifically, even within the CF community itself the information circulating about aging with the disease often proves inconsistent and at times contradictory. Beyond the experiences of adults with CF ourselves, these misconceptions are magnified and entrenched. Conversely, the process of more adults with CF surviving to older ages is constantly providing concrete and diverse data to combat both active misinformation and passive lack of knowledge concerning the experiences of people aging with the disease.

An important means of building on the findings from this autoethnographic inquiry will thus be diverse approaches to scholarly engagement of representations of aging with CF—both those generated by patients and those created by others in their lives. I learned from my own research on my personal lived experiences that the perspectives of both other aging CF patients and people without the disease yield unique value for my own understanding of what it means for me to grow older. These same inputs have enriched my broader sense of what may be possible in the future for people aging with CF as a population. At the same time, conducting critical autoethnography on my own personal history of aging with CF has heightened my sense of caution about attempting to generalize my experiences beyond the context of my own life. This manuscript is best interpreted as both a detail-rich examination of the trajectory of one patient-scholar aging with CF and a context-specific reflection that encourages similar critical thinking about other people's narratives of living and aging with the same disease.

As I have gained insight into what aging with CF means for me, I have felt more capable and competent in articulating my hopes, fears, and goals (see Corbett, Foster, & Ong, 2007) about the future to both my care team and my loved ones. I have received similar feedback from other adults with CF about their experiences. Indeed, I have facilitated this process in real time myself by engaging in advocacy and outreach activities with the CF Foundation and other groups. I have learned that although healthy aging with CF remains a massive and complex challenge, it is also an exciting frontier on which new developments are constantly changing our sense of what growing older with this disease can and should look like.

My explorations with this autoethnography suggest that as a society, we can best improve our collective awareness and understanding of aging with CF by sharing our stories, listening to those of others, amplifying these narratives, and reflecting on the inclusiveness of our discourse. This process has already begun in diverse ways within the CF community, from social connections forged in patient online communities to advocacy efforts to creative projects. But these voices have not yet found purchase in conceptualizations of aging with CF

beyond the community itself and those closest to it—patients, our families, and our close friends. Diversifying and refining popular consciousness concerning aging with CF will require much more visible representations of adults with the disease in mainstream media.

I will thus conclude with an anecdote about the power of television. While doing research and writing field notes for this autoethnography, I started watching *Bates Motel* on Netflix. Not knowing much about the show or how its storyline would differ from the original *Psycho* film by Alfred Hitchcock, I was astonished to see a young adult with an oxygen tank walk onscreen. As my brain immediately leapt to wondering if this character (Emma Decody) had CF, my question was quickly answered in the affirmative as she explained to the other central characters why she had to use supplemental oxygen. In that moment I felt astonished and overjoyed to see a main character with CF on a popular television show, and realized quite acutely in that moment how much I had been missing in my life by never seeing representations of my disease in mainstream media apart from “cases” on medical shows—more numbers on charts and puzzles to be solved than people with complex lives and personalities.

Yet I also looked at my own thinking with suspicion, specifically with respect to my immediate questioning of whether Emma had CF. I would like to think that my desperation to see any representation of my own lived experience on television—even one that does not closely mirror my own personal trajectory with the disease—lay at the root of my brain clinging to the idea that she might possibly share that fundamental characteristic with me. However, I also wondered if I had jumped to that possibility inappropriately. Certainly, there are other conditions that can cause a young adult to require supplemental oxygen. Likewise, I have spent much of this manuscript cautioning against assuming that a given person aging with CF will necessarily require oxygen or other specific interventions at any point in time.

Ultimately, following Emma’s story on *Bates Motel* proved very instructive and affirming. Her story was the farthest thing from the kind of “sadness porn” I described in my early rants to my family about popular depictions of CF. Emma was adventurous, resilient, and determined. Eventually she got listed for a transplant and was able to stop using supplemental oxygen after a successful surgery. Seeing her go through that when I was facing the possibility of needing a kidney transplant in the future—a reality that still lurks at the fringes of my consciousness on a daily basis—gave me the courage to join the online CF community myself and open my mind to the transplant stories I knew I would hear. It gave me a sense that even if my own process of aging with CF included transplantation, I could still be a “success story” (see Ouwehand, de Ridder, & Bensing, 2007). Moreover, it challenged me to rethink what “successful aging” means to me overall as a person with CF (see Minkler & Fadem, 2002).

As I formed friendships and collaborated on outreach activities with other adults with CF—both those with stories like mine and those with very different experiences—I began to answer that question. I am still answering it today and suspect that I will be reflecting upon it every day until I finally do reach the end of my life. One of my goals now, which would have seemed ridiculous even a year ago, is to be the oldest living person with CF when I do finally expire. Whether I actually reach that goal matters little in comparison to the fact that I give myself the freedom to consider it, and to plan for a future that includes being old. For an aging researcher who spends most of their days immersed in scholarship on later life, it was a special kind of torture to contemplate the appearance of my own days being severely numbered.

Thinking about the future with a more open mind has presented its own challenges—foremost among them a massive amount of cognitive dissonance and a frequent feeling of being hopelessly lost (see Brashers et al., 1999). Yet in this process I have come to accept a different kind of ambiguity (Mishel, 1999) in my life, the kind that makes us ask ourselves, “What will I be like when I’m older”? I cannot answer that question yet and will not be able to do so conclusively for quite some time. But replacing “if” with “when” in that eternal question to myself represents a major step forwards in my thinking about my journey through life as a

person with CF. As for what I actually will be like when I do reach old age, and how I will feel about all of it...that will simply have to become the subject for a future autoethnography.

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