Healthcare Transition from the AYASHCN's Perspective

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When an Illness Is Life-Limiting

Looking forward to a future is a prerequisite in health-care transitioning. Transition starts at diagnosis, as parents start to imagine the future. When Ana and I were first diagnosed, my parents began to grieve and cope with fears about our early deaths. My mother expressed feeling overwhelmed by the caregiving burdens and not being able to even imagine how we'd handle this as we grew up. Over time, my parents felt more competent in managing our treatments and focused on ways to control aspects of the illness. Fear of our early deaths diminished, but what remained motivated our family to live a normal and positive life as possible, not knowing how long it would last.

I learned about the possibility of my mortality at 10, when a hospital friend with CF died. The anxiety and fear of dying drove me to do take my health needs seriously, so I could have a future. With much adherence and effort, I outlived my prognosis. This accomplishment fueled my sense of agency.

The most important word for those with life-limiting conditions is *possibility*. I wish someone had talked to me when I was an anxious teenager and told me to believe in all of the possibilities of my life. Yes, dying young was a real possibility. Yet growing up, going to college, falling in love, getting married, getting a lung transplant, and having a career were also all possibilities. I just had to work very, very hard doing my medical treatments and have some hope, grace, and luck.

One of my friends with CF explained our situation as follows:

> *You are planning on taking two trips: one to the North Pole, and one to Hawaii. You have to pack your suitcases to be prepared for both climates. So pack your down jacket and your bikini. In other words, plan your life journey with the possibility of living a full life, and plan your other journey with the possibility of death. We learn to live and cope with challenges imposed by illness with full awareness that "anything could happen."*

This philosophy allowed me to live in both denial and confrontation of my reality. If we prepare to take ownership of our medical care as children and teens, then we will not be caught off guard when we survive. We will be confident in managing our health needs as we pursue adulthood. What a celebration! If, sadly, we do in fact die, we have embraced life with hope and optimism, which enhances the quality of our lives.

HCT as a Normative Event

It is normal for all children to move from preschool to high school as they grow. We become comfortable with a classroom of friends, but inevitably, we move on to the next class and meet new teachers and peers. We say goodbye, gain trust,

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and become settled in the next classroom. It is a normal developmental process, then, for adolescents and young adults with special health-care needs (AYASHCN) to move from a pediatric health-care team, one with whom we feel familiar and comfortable, to an adult care team who can care for our childhood illnesses and also better address adult health-care needs. It *is* important for adult patients to see providers trained in adult medicine. A tragic example of this is my sister Ana's extreme gastrointestinal pain that developed in her late 30s. A pediatrician may have attributed this to normal CF issues, but sadly, she was ultimately diagnosed with colon cancer: a curse that seems to afflict older adults with CF.

Parents naturally teach their children to take on more responsibility as they grow; each new task mastered fosters self-esteem. My parents had the wisdom to pair normative developmental tasks with illness-related tasks. For example, when I was old enough to brush my teeth without a reminder, I was old enough to take my pancreatic enzymes at meals by myself. When I was old enough to call a friend on the phone, I was old enough to order my medication. When I was old enough to wash dishes, I was old enough to sterilize my nebulizers. When I was old enough to drive on freeways, I was old enough to drive to medical appointments by myself. With each success, my parents learned to trust me; with each mistake, they learned to supervise a bit more. This is typical for all parenting.

Moving away from home is a normal rite of passage. In high school, like all my friends, my great focus was getting into a good college. Then Ana and I were accepted into a university 400 miles from home. Living away from my parents, then, meant that Ana and I would be completely responsible for our health-care needs. By 18, we had to be prepared.

Promoting Autonomy in Disease Management by Family and Health-Care Providers

Every parent's goal is to raise their child to become self-sufficient, to make smart decisions for their lives, and to be happy. Learning to gain autonomy with a health condition can be an *opportunity* to facilitate growth. AYASHCN have the potential to learn resilient traits such as accelerated maturity, taking responsibility, acceptance of prognosis, gaining control, redefining normality, finding social support, and enhancing problem-solving [1]. These traits can foster success in academics, sports (when appropriate), and hobbies and eventually in a work environment.

Autonomy in disease management is gained gradually, as parents, teachers, friends, and health-care providers all participate in supporting a child's learning. Promoting autonomy starts with the family. It is not unusual for parents to feel protective, guilty, and responsible for their ill child's survival [2]. Without any choice, a parent (usually a mother) becomes a case manager, nurse, and expert in her child's disease. The parent retains the power in the family when it comes to medical care. Sometimes, this mother can struggle to relinquish this power to the patient. I have met a 40-year-old with a congenital heart defect whose mother still put her pills out for her. Recently, a mother of a teenager with CF shared:

I'm so scared that when my son leaves for college, he'll undo all the hard work I've put into raising him to be as healthy as he is, and I'll have no control over him.

When a teen leaves the home, the parents face the loss of a deeply dependent relationship, loss of their role, and loss of purpose. I had friends who became nonadherent in college and their health deteriorated permanently; they had to learn the hard way why treatments were important. Relinquishing control is one of the hardest things a parent of an AYASHCN can do. I met a mother of a CF child who exclaimed to me her epiphany:

I finally realized my daughter was the one with CF, and that I didn't have the disease. It was up to her to learn how to live with her CF. I could do everything I could to manage her CF, but ultimately she's the one who has to manage her CF.

As a young child, I had many opportunities to practice owning my health care. I remember the first time I had to go to the nurses' station to take my pills, around kindergarten. I was self-conscious and angry that I had to be singled out. Eventually, I exerted my autonomy by either skipping these pills or by fourth grade, I packed my own pills in my lunch bag so I could take them myself.

Sleepovers at friends' homes were other opportunities to manage my health needs away from my parents. My mother allowed us to have an active role in the decision-making process: did we want to do our treatment before we went to Jenny's house, or did we want to do our treatment at Jenny's house, with Jenny? By 10 and 12, respectively, my mother sent Ana and me off to spend a week with our grandmothers. My parents sent a letter of instruction to our grandmothers about our treatments and instructed us to "teach" our grandmothers about what we needed for our therapies. My mother packed our pills and told these surrogate parents to make sure we took them. This open communication helped to ensure that all parties were informed about what needed to be done for us to stay healthy.

Another milestone was when I spent the night in the hospital by myself for the first time. I was 5, and my mom said she had to go home to take care of my sister and brother. I remember being terrified. Yet, I learned to use the call button and ask the nurse for the bedpan. Today, many children's hospitals have a bed for a parent in the child's room. The parent provides comfort and familiarity. However, this also sends a message that the child needs a spokesperson and isn't safe alone with only nurses to care for her.

Establishing the habit of adherence was essential to teach autonomy. My parents taught us that treatments were nonnegotiable. When we were 5, my dad told us, "Every treatment you miss is one day less of your life." He drilled into us the importance of choice and consequence. Also, my father installed a battery in our family van, so we could do treatments on the road. Adherence required creativity. As teens, my parents joked that we could get tattoos and piercings, dye our hair, or do anything else to our bodies, but we could not skip our treatments.

My mother became a respiratory therapist when my sister and I were in grade school. As a working mom now, she needed us to help out with housework as well as our own treatments. As twins, Ana and I encouraged each other to be compliant. We'd compete over who could tolerate longer chest percussion or who'd cough up more mucus. During high school, my older brother drove us in the family van to school, and we'd go to the van each lunch break and do a treatment.

Health-care providers also fostered our autonomy. As teens, our pediatrician drew pictures of lung alveoli to describe atelectasis and other disease processes, so we became knowledgeable about our disease. We learned why we needed our treatments and IV antibiotic "tune-ups." Education motivated us.

Ana and I learned to become self-reliant from disappointments as well. While hospitalized, we often pushed the call button when the IV machine was beeping. Unfortunately, sometimes it took over 30 minutes for a nurse to respond. Anxious about clotting my IV, I learned to turn the alarm off and release saline to flush my line. Frustration led to problem-solving and autonomy.

Most AYASHCN do not have a sibling who shares the same health challenges. Ana and I developed an enmeshed relationship, which allowed us to separate from our parents. In an effort to spare our parents from the burden of our care, we started to do chest percussion for each other at age 12. We took charge of our treatments when we moved away to college. When Ana and I were seniors in college, our clinic coordinator started to assign different exam rooms for each of us. At first, we were resistant and angry. The coordinator said we needed to differentiate and learn to speak up for ourselves, without the crutch of the other twin in the room. This was the first step in helping us gain true autonomy from each other.

The Role of Parents

Parents create a family culture, with unspoken or spoken themes [3]. Themes might include empowerment, anger, victimization, blaming, overprotectiveness, or enmeshment. These themes will influence how a child learns to own her illness and how the child ultimately transitions to independence.

In my family, themes such as striving for normalcy, yearning for control, and self-sufficiency helped us develop self-esteem based on what was normal: our studies, our love of nature, creativity, pets, and friends. These "normal" things gave our

lives meaning and purpose. They helped to define my identity. My parents had "The Serenity Prayer" framed on the wall at home; they believed in controlling as much as they could around our health and surrendering the rest. We were constantly planning, problem-solving, and strategizing how to maintain our health: from doing up to five treatments a day to researching clinical trials and to asking our doctor for new medical devices. We incorporated treatments into our daily routine: early in the morning, we'd head to our parents' bedroom for therapy, and after dinner we'd lay over their laps in the living room during the nightly news for chest percussion. My father's German and mother's Japanese cultures valued self-sufficiency. When we missed weeks of school, my parents didn't believe in IEPs (Individual Education Plans). They encouraged us to call our friends for the homework assignment, and my father tutored us in math and science.

My parents expressed gratitude that we had so many opportunities and such good medical care to take care of ourselves. My parents believed in "no pain, no gain" and that raising us took sacrifice. Each lunch hour in junior high school, my father would drive 20 minutes to our school parking lot to help us do a treatment. He passed this duty onto my brother when we made it to high school.

And because no family is perfect, my mother was plagued with depression when we were teens, mostly caused by marital distress and our worsening health. She would work, come home, and go to bed. This forced us kids to make dinner, pack our own lunches, and initiate treatments on our own. Inadvertently, she was preparing us to take responsibility. Ultimately, she worked through her depression.

Though my mother experienced anticipatory grief over losing us, some denial guided her to teach us to become self-sufficient. When Ana and I were 12, my mother taught us the tedious task of washing our nebulizers every night. I used to cry and plead for her to do it for me. While it was hard for her to see me upset, she did not give in. She insisted this was a chore that was required when I grew up. This chore expanded to include learning how to clean a toilet, do laundry, and so on, so I could function as a young adult living on my own. I had no privileges as a sick child!

When I left home for college, I remember my father's stern command, "Enjoy your time at Stanford. Remember, first comes health. Then comes your studies. Then comes everything else." My mother's worries led her to call a cystic fibrosis nonprofit agency close to our college to see what kind of support they could offer us. Soon, we were invited to homes of CF families and developed a close-knit community of friends. This relieved my mother's fears tremendously.

Shortly after we arrived at college, we attended a cystic fibrosis specialist for the first time. When the doctor reviewed our lung capacities and BMI, she stated, "I can't believe your parents would send such sick daughters away from home for college!" We were taken aback by her comment but also knew that my parents believed that in some way quality of life was as important as quantity.

My medications were all changed at my new clinic at college. Soon, my parents no longer knew what meds I was on. Throughout my young adult years, when my mother would call us, her questions always focused on health: did we do our treatments, how were we feeling, how much sleep did we get, what medications are helping, and so on. Over the years, I felt my parents didn't really know about *my life*, except that I was alive. I now encourage young parents I meet to ask about their child's interests, friends, and what they did lately, before they ask about health.

As independent as my sister and I were in college, we never fully gained independence from our parents, until we got married or started working. We remained on our parents' insurance under the medically disabled dependent program. My parents would visit when one of us was hospitalized. They helped us financially throughout college and graduate school, because we didn't have the energy to work part time and earn extra income. Every school break, we'd return home and resume having our parents help us with treatments. My parents assured us that they would always be there and that we could even return home if our health worsened. This could be a reality for many AYASHCN.

The Role of Peers/Support Groups

It is normal for adolescents to shift their attachment focus from parents to peers. A transformative event in my life was attending cystic fibrosis summer camp. At 11, I learned there were children like me, all facing the burden of learning how to manage their medical care. At camp, we learned that there were trusted adult counselors who could do treatments for us and supervise our medications. The education at camp helped me gain mastery over my disease. I also saw kids with CF who didn't do their treatments, and over the years those kids didn't return. We held annual memorials for the kids who died the prior year. This reality motivated me more than anything else that in order to survive, I *had* to be adherent. Camp also exposed me to older adults with CF who took their treatments seriously. They became my role models and showed me the positive possibilities for my future.

CF camp taught me how to accept CF as part of who I was. Owning my CF and incorporating CF into my identity helped me speak up for my needs for the rest of my life. If I resisted CF, denied or dismissed it, I could never fully advocate for myself. CF camp made me feel special rather than different. Some good things came out of having this disease. My CF peers helped me let go of shame.

My many months in the hospital exposed me to other sick children who became my friends. I met kids who had diabetes, cancer, kidney disease, and sickle cell anemia. I learned that other kids have to deal with different challenges. But I also saw kids who didn't take care of themselves and overheard many angry lectures from their parents and health-care providers.

Like any teenager, I had a strong need to fit in and belong. While I fit in academically with my healthy peers, my frequent absences, violent coughing episodes, and being a late bloomer made me painfully self-conscious. Yet, my illness became a weeding tool, and I attracted compassionate friends who could understand my medical needs. They asked questions and showed me that I didn't have to waste energy hiding my medication or holding in my cough. Some visited me in the hospital. My friends would help with chest percussion or ward off smokers at the mall. As we entered adulthood, friends expressed gratitude that Ana and I taught them about appreciating life. They helped me practice telling others about my CF and trusting that some people are capable of embracing it. Eventually, I tested this practice with dating and was lucky to find a husband who could handle my health needs and love me for who I am. I have been happily married for 19 years.

In summary, providers can encourage families to get involved with disease communities and camps. Hospitals can provide support groups for families to create a safe place to learn to own the disease. Online peer-to-peer support for digitally minded teens and young adults can be invaluable. Providers can educate children and their parents how to speak openly about their health condition to peers and schools. Positive relationships are healing and life prolonging.

Promoting Self-Efficacy with Treatments

Self-efficacy is the extent or strength of one's belief in one's own ability to complete tasks and reach goals. Self-efficacy begins at a young age. As a child, I discovered that if I didn't take my enzymes, I would have a stomachache. My brother would make fun of the smell in the bathroom. That was enough to instill the habit of taking my enzymes.

As my disease progressed, I was overcome with fear of dying. I developed an anxiety disorder that remained unrecognized and untreated. My parents were emotionally ill-equipped to help me cope. Eventually, I harnessed my anxiety to motivate me to do more treatments religiously. Ultimately, increasing the frequency of treatments helped me stay out of the hospital. I felt empowered that I could "overcome" CF. New antibiotics and medications became available that helped me stay well. Then my drive to keep up with treatments was fueled by the hope that new medications and a cure would be around the corner. We joined a clinical trial as young adults,

and this dramatically improved our lung capacity. Clearly, self-efficacy is cultivated when there are positive results from our efforts.

Advice for Parents

Many parents of CF children have asked me when to tell their child about the life-threatening nature of the disease. They fear that if a child learns she can die of the disease, she will lose hope and grow anxious. A child knows innately that their condition is serious. And with today's digital world, it's easy to find out the hard way about a prognosis online. I am an advocate of offering honest, developmentally appropriate information to children. Generalizing human vulnerability by saying, *We all will die someday. That's why I eat healthy and exercise. Just like doing your treatments, taking care of myself will let me be strong for as long as possible*. Offering an outlet for emotions, such as a support group, camp, or therapist, or having family discussions about how the child feels about her disease and her future, is important.

Setting goals and fostering a sense of self-worth will help motivate AYASHCN to live with optimism and hope. Unaddressed depression and anxiety will impact adherence [4]. Including healthy siblings is important, because the prognosis will impact them as well. My brother worried about losing his sisters, told the neighbors we were dying, and turned to drugs and alcohol as a teen to cope. Ultimately, offering supportive resources to AYASHCN to promote resilience is as important as medication itself. With this support, the child will find her own way to learn to regulate her own emotions around the disease.

School

School is the most normal experience of childhood; it exposes kids to academics, sports, music, and clubs that provide the foundation for self-esteem and self-efficacy. My parents encouraged my sister and me to focus on what *was* working. School gave us the chance to be good at something, which allowed our identities to encompass more than just being sick kids. Our teachers encouraged us and praised our academic achievements. Some teachers came to our hospital room to administer tests. Our academic successes played a significant role in helping us feel respected, responsible, and capable of self-efficacy.

There can be a limit to self-efficacy. As our disease progressed, we increased the frequency and duration of our treatments. Sometimes our hands bled from doing hours of chest percussion. When my sister started working at a hospital, she coordinated with a respiratory therapist to give her chest percussion during lunch break. But eventually, the striving to *do more* no longer offered positive outcomes. We felt like failures. My parents' motto rang true: "Do your best, that's all you can do." We finally acknowledged what a cruel disease we had.

Promoting Self-Advocacy

One benefit of having a chronic condition is learning how to express one's thoughts and feelings. This is the underpinning of self-advocacy. As I grew older, I gained body awareness and could read the slightest change in symptoms. I learned to articulate any concerns to my providers. By 15, my doctor met me alone with my sister in the exam room, while my mother waited outside. He would ask what was *really* going on. Of course, we hated to be hospitalized and wished we could pretend all was well. But our clinical numbers (lung function, weight loss) were the evidence. My mother would join at the end of the visit to affirm our reports and express her concerns but only after we had the chance to discuss ours.

By early adulthood, my illness, as well as my health-care providers, taught me how to express basic feelings: "I think, I feel, I need, I want…." This was a breakthrough for me. My mother's Japanese culture taught me to not burden others and to be passive. But this approach would shorten my life. If I didn't speak up for my health needs, no one else would. Even my twin couldn't feel what I was feeling; she couldn't speak up for me.

Self-advocacy starts at an early age. Learning how to tell people the name of your disease and what it affects is a milestone. This empowerment means the child is no longer a victim but an educator and ambassador for her disease. A number of my younger friends with CF did "show and tell" in grade school, sharing about their illness. A vibrating vest has a coolness factor when classmates can try on the device. It takes practice to become accustomed to saying, "I have [this condition] and that's why I need to do [my inhaler before soccer practice]." In some cases, a fundraiser like a walk is an opportunity for kids to speak up to their peers, neighbors, and friends, as a conversation starter about their illness. I remember overcoming my embarrassment and inviting my friends to join a fundraiser called "Bowl for Breath." They enthusiastically joined. Telling others about your illness is a requirement in adulthood, when we need to request accommodation in the workplace, health care, housing, or travel.

Self-advocacy often develops in response to emotions. As a teen, I would get angry when nurses came into my hospital room asking when did we "catch" CF (CF is genetic). One resident said, "You have CF? At your age, you're supposed to be dead!" We faced much ignorance and carried tremendous anger. I felt a strong desire to educate everyone about CF. Fear also fueled my self-advocacy. As a young adult, I was scared of being disliked by my providers. I thought that if I upset them, they wouldn't take good care of me. As a teenager, a nurse was going to inject an antibiotic into my IV line. She dropped the syringe, without the cap on, picked it up, and prepared to insert it into my line. Terrified of infection, I exclaimed, "No! Please get another needle!" I made the nurse upset, but my fear fueled my outspokenness. Similarly, I was terrified of multiple needle sticks when the nurse had to start an IV. It took time to learn to be assertive to tell the nurse what vein to use.

This advocacy continued in college, when we visited the Disabled Students Services office. It was a relief that our college offered accommodations. Ana and I asked to room together, and for a private bathroom in the dorm room, to avoid infections. We overcame shame and joined the Disabled Students Speaker's Bureau to educate students about living with a serious illness. As much as we wanted to be normal, having CF was *not* normal, and we had to accept this and advocate for special services.

Readiness to Transition

All children have their own pace in transitioning. I do not believe there should be an absolute age for transitioning, such as 21 years. Factors that may impact transitioning include emotional maturity, intellectual functioning, disease awareness and acceptance, family dynamics, and self-esteem. I've met 25-year-olds with CF who would rather play video games than do their treatments; and I've met 16-year-olds with CF who are fully independent, take their high school equivalency test, and start working. Culture matters too; my parents came from countries where children earned independence at a younger age than in America. In Japan, it is not uncommon for 6-year-olds to take the train by themselves. So, by 8, my mother encouraged us to walk to school, to take the bus, and to learn how to manage our medication at school by ourselves. And because of my mother's accent and English difficulties, she asked us to speak up for ourselves and for her at a young age.

Socioeconomic status (SES) can ease the transition to independence. Ana and I were privileged; at 18, my uncle bought Ana and me a car to share. This gave us the freedom to go to the doctor by ourselves. SES can impact going away for college. AYASHCN may have to continue living with their parent for longer than their healthy peers. Some CF friends had to return home after college because of inability to work full time. Medical expenses can impose significant financial burden for many AYASHCN. It's important to recognize that "success" as an AYASHCN does not mean true independence from family members. AYASHCN should always be validated, encouraged, and praised for any incremental steps toward self-reliance.

The physical condition of the patient impacts the ability to transition. It can be difficult when a

parent sends a sick teen away to college, and their health deteriorates significantly. I have lost several friends to CF during college. The parents were racked with guilt for letting their child go despite severe illness. Over time, they could make peace with the fact that they allowed their child to pursue their dreams, despite the outcome, and the young adult truly lived.

When my sister was 24, we were encouraged to see an adult provider. At this time, her lung capacity was roughly 25% and she was evaluated for a lung transplant. She felt abandoned by her pediatric team, and by sending her off to an inexperienced adult pulmonologist, she shared, "They were giving up on me." It would've been more appropriate to wait until after her transplant to transition to an adult provider or transition earlier like age 21 when her health was still stable. It is not appropriate to transition a patient when they are in crisis.

Addressing Mental Health, Substance Use, and Reproductive Health Issues

I appreciated having *normal* teen and young adult issues addressed by my providers. I hated when they only cared about my chest! My illness impacted me physically and emotionally, so having providers who took my mental health seriously was important to me.

My mental health was not addressed by my pediatrician, though he provided words of encouragement and a caring presence. The inpatient social worker spent the time to ask Ana and I how we were coping with our illness. We opened up to her, more than to our parents. She helped us see that we could hate CF but didn't have to hate ourselves. Our social worker also counseled my mother through her depression, and I was glad my mom was getting support. Much later, I learned that the number one predictor of a child's coping with their illness is the coping skills of the parents [5].

After college, the social worker continued to be our primary mental health resource. It helped to learn how certain medications worsened anxiety and tools to cope with anxiety. When my sister reached end stage and needed a transplant, I grieved losing her as a sister and caregiver—along with my own declining health. Both of us cried at nearly every clinic visit. My doctor prescribed an antidepressant in my late 20s. There was no stigma; most of my CF (and many non-CF) peers were using antidepressants. And my social worker suggested I see a psychotherapist. Writing a memoir became a therapeutic outlet as well.

My peers with CF impacted my mental health significantly. They taught me to *be stronger than your illness*. They taught me the motto, *Do your 'I hates' first.* Endure what's uncomfortable, so you feel accomplished afterward. However, the cost of belonging was witnessing numerous losses. One after another, our CF friends died. The grief exacerbated my anxiety and depression.

Rebellion is one way to deal with the intense emotions associated with illness. Rebellion can lead to self-loathing, unreasonable anger at others, and nonadherence. Providers can help AYASHCN find healthy ways to rebel against their situations. What helped me was the use of profanity and body art, expressing myself in art and writing, my spirituality, complaining to people closest to me, and directing my frustrations toward healthy distractions.

It was part of a routine psychosocial assessment to ask about use of drugs, alcohol, smoking, or other rebellious behaviors. My providers made no assumptions about how I looked or what I was like and asked about these behaviors. My providers educated me on the dangers of alcohol interacting with some medication. I witnessed plenty of substance use in my CF peers. I never got involved in drinking, smoking, or drugs, because I valued an intact mind and didn't have the luxury of experimenting with anything that hurt my body. For some, getting drunk was a way to escape CF, at least for a bit. Their friends used substances, so they wanted to fit in. Soon, these behaviors became a habit. And for some, their habits shortened their life or made them ineligible for transplantation.

Since I was a teenager, my providers also assessed my sexual development and activity. Since I had no development until my late teen

years, I wasn't shown the embarrassing Tanner stage pictures until I was around 17. I had terrible body image and couldn't imagine anyone wanting to date me. I was 22 when I met my first boyfriend. I was put on birth control shortly after I started dating, to regulate my periods, but also for other benefits! Right after my wedding to this boyfriend, I lost my parents' insurance and was now on my husband's insurance. The first thing my new adult doctor asked about was whether I was planning to get pregnant. He then told me that pregnancy could exacerbate my lung condition, so I (and my husband) accepted that we'd be childless. In sum, it's important to not make any assumptions about AYASHCN. It is a myth in the health-care world that young people with serious health issues are asexual. I knew a friend with 11% lung capacity who was still having sex! Sexuality is an outlet for pleasure and a way to befriend even an ill body.

Finding an Adult Provider

Access to competent adult providers is critical for all AYASHCN. My sister and I were part of the early generation where adult CF care providers were not yet established. We visited and fired a number of adult providers who, while well-intentioned, did not have training on how to care for us. Several said CF patients are knowledgeable and can tell them what they needed. True for me, but I still needed a doctor.

The week we drove to college for freshmen orientation, my mother joined us to visit a local adult pulmonologist. This doctor introduced herself by asking, "So, how can I help you?" We were turned off by her lack of acknowledgment that we had long medical history and required significant medical guidance. This clinic was not a Cystic Fibrosis Center, accredited by the Cystic Fibrosis Foundation. This adult pulmonologist had not heard of the national CF conference for providers. I found a reliable pediatric care team much farther away and stayed with them through college until my mid-20s.

This pediatric team developed an "adult program" incrementally. Initially, the core pediatric team joined one adult pulmonologist. This gradual transition offered the familiarity of my pediatric team with the introduction of a new adult pulmonologist. The team reassured Ana and me that this doctor was competent. By then, I was the expert of my own body. But I was struggling with new issues like bowel obstructions, diabetes, and massive hemoptysis and was scared. I needed someone who could care for increasingly complicated disease.

Part of my gradual transition meant that when I needed to be hospitalized, I was admitted to the adult hospital. This is when I missed the pediatric nurses the most; they were friendlier than the adult providers. Hand-holding didn't exist in the big adult hospital. Over time, both CF care teams worked hard to train the staff to provide the kind of care that CF patients were used to in the pediatric system. Even adult gynecologists, endocrinologists, gastroenterologists, lab technicians, RTs and PTs, and primary care doctors were trained to care for CF adults.

It took almost a decade after I moved to college to find a fully functional CF adult care team. My pediatric team coordinated the transfer of care so the adult team knew my history before I showed up at clinic. I appreciated having an adult-trained nurse, social worker, nutritionist, respiratory therapist, and internist/pulmonologist devoted to all of my care needs. The entire team attended the national CF conference, making me feel confident that they knew what they were doing. But working with young adults within a family system took practice. Family boundaries are important for adults. One of my CF friends in college shared that her adult doctor called her mother back home to see if this student was taking her medication, since her lung capacity was declining. My friend angrily confronted this doctor. It's important that providers treat their patients as adults and respect their confidentiality unless explicit permission was given to share their health information with their parents.

Nowadays, there are more systematic processes for transitioning. At specific ages, young patients are educated in advance about transition goals and are expected to meet those goals [6]. Young adults are introduced to their adult providers ahead of

transition, with the familiar pediatric team present. This meet-and-greet opportunity breaks the ice and gives the patient a chance to interview the provider and get a sense of their personality and expertise.

Terminating the Relationship with the Pediatrician

Saying goodbye is one of life's most normal and yet difficult rituals. Termination with our pediatrician happened organically, when we moved away for college. For a year or two, during school break we'd return to our pediatrician for a "tune-up." Gradually, Kaiser developed a coordinated CF Center, and we were transferred to the new program. We kept in touch with Christmas cards until my pediatrician died 3 years ago. I never imagined I'd outlive him. I will never forget how my pediatrician encouraged Ana and me to lead a normal life and apply for college, despite our advanced disease. He told us, "Follow your dreams." I will forever be grateful for his blessing.

The pediatric team who cared for Ana and me during our young adult years became a family we never wanted but learned to love. They witnessed the toughest years of end-stage disease, supporting us through many tears and triumphs. They saw my sister through her transplant.

When it came time for my transplant, I had already transferred to the adult team. I deteriorated into respiratory failure and was intubated. No one was sure if lungs would become available. I was touched to hear later that my pediatric team came to my bedside, while I was sedated, to say goodbye. They *really* cared. And then, when lungs were offered and I survived, they all returned to celebrate with me.

In summary, one piece of wisdom I've learned in 45 years is to question the American value of "independence." An AYASHCN will feel like a failure if she can never achieve the desired independence. But no human being is truly independent. We are all *interdependent*. It really does take a village to raise a young person, especially an AYASHCN. From parents to teachers, to counselors, to friends, to an illness community, and to the pediatric and adult care teams, all work together to guide and encourage self-management in the young patient. The goal of transitioning should not be independence, but developmentally appropriate self-management of health-care needs. AYASHCAN should be encouraged to find ways to feel satisfied with a sense of independence within a dependent situation with peers, partners, and, in some cases, still parents. No matter what, leaving a pediatric care team is a coming of age experience and a cause for celebration. This transition is part of a success story.

Isabel dedicates this chapter to the memory of her twin Anabel Stenzel, whose reciprocal caregiving allowed her to transition and survive to middle age, while sharing an extraordinary life. For more about the Stenzel twins' story, see The Power of Two: A Twin Triumph over Cystic Fibrosis (Missouri, 2014) and their documentary film, www.thepoweroftwomovie.com.

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