Acceptance & Commitment Therapy for FSHD

A psychologist's personal journey and clinical guide

By **Kent Drescher, PhD**

Acceptance and Commitment Therapy for Facioscapulohumeral Muscular Dystrophy (FSHD):

A psychologist's personal journey and clinical guide.

by

Kent Drescher, PhD

Copyright

Acceptance and Commitment Therapy for Facioscapulohumeral Muscular Dystrophy (FSHD): A psychologist’s personal journey and clinical guide.

www.kentdrescher.com

ISBN: 978-1-7376758-5-3 (PDF format)

Disclaimer

This book is a self published work. As a result I have no one to blame except myself for errors that you may find inside. Because of my FSHD, I use voice recognition software to write. This software is extremely helpful, and almost never makes spelling errors or typos. However it does tend to make ‘sounded like’ errors. These kinds of errors don’t always show up with standard spell checkers, and close proofreading is required to catch them. Several skilled and knowledgeable people have read the book and provided comments and suggested edits. A skilled proofreader has also been through the text. All that being said, it is likely that some of you will still find mistakes of varying types in the text. If you do please reach out to me and I will fix them in the next revision.

The ACT community is large, and I am a relative newcomer. I’ve made every effort to give proper credit to the originators of the various ideas, metaphors, and exercises created by others, from which I’ve borrowed and modified for FSHD. If I’ve done this incorrectly or missed something, please know it is entirely unintentional, and I apologize. Please reach out to me and let me know, and I will endeavor to make corrections and give proper credit.

Dedication

I am so grateful to have a loving and supportive family that has walked with me each step of the way on my FSHD journey. I am also thankful for the caring and support of close friends and colleagues. I am so lucky to have Robyn Walser, PhD, as a close friend and supporter of this effort. She is a world renowned expert in Acceptance and Commitment Therapy (ACT). Much that I have learned about ACT, she modeled by her actions and instruction.

I am grateful for the FSHD research community, a group that works persistently toward a cure for this terrible disease. I appreciate Dr. Christopher Graham. His work with ACT, which includes a focus on a range of neuromuscular diseases, inspired my interest to pursue this book and assist others with FSHD. His work also motivated me in relation to my personal struggle with FSHD in a fruitful and values-based direction.

I offer thanks for the efforts of the FSHD Society, a patient advocacy organization working tirelessly to bring patients together with research labs, pharmaceutical companies, government agencies, and with one another in marching forward together in search of a cure.

Ultimately, I dedicate this book to all those who suffer bravely from FSHD and to their families who are part of the journey. They each deserve so much more!

Forward

Finding Meaning in a World Not Chosen

By Robyn D Walser, PhD

When I was 11 years old, my younger brother was diagnosed with Legg-Calve Perthes Disease. This is a disease often diagnosed in childhood. The blood circulation to the hip joint is blocked, and the bone at the femoral head begins to die, destroying the round end of the femur, the "ball" that sits at the top of the bone and fits into the hip socket. Several treatments are designed to keep the ball snuggly within the socket so it doesn't weaken and begin to fragment. This is where Bobby's journey (as well as the family's) of emotion, thought, and sensation challenges began. We did not choose this journey. Still, it was there to be made.

Bobby was first placed in a brace designed to keep the ball "held" in place. A bit unusual, it consisted of a one-inch round metal bar about three feet long with two inward-facing orthotic shoes attached to each end. When wearing the brace, Bobby's legs and brace formed a triangle, with his feet held out constantly at a three-foot spread and turned inward. He had to wear the brace except when he was sleeping. Moving through the house and environment as well as adjusting to keeping his legs fixed in this position, was emotionally challenging. He was captured - forced to hold his legs in this awkward and uncomfortable position.

Despite this challenge, Bobby did an interesting thing. Rather than trying to walk with his legs spread at such a wide angle, he adapted. He discovered that he could move more quickly and maneuver more easily if he placed both hands on the floor and walked on all "fours." He essentially turned himself into a kind of quad-pod. He would bend over, scoot through the house, and move around using his hands, and a kind of waddle with his feet. He became quite good at this way of moving and had the family in stitches with laughter at different times through this months-long experience. Don't get me wrong, there were also many tears and times of heartache for him and the family. Seeing my little brother trapped in this contraption was hard and painful…and it didn't work.

After six months, the family learned that the brace had failed. Bobby's hip was not improving. The blood flow was still not getting to the bone. The next step meant even less movement. Bobby was fitted with a small wheelchair and confined to it during the day. The doctor hoped that holding the hip still in this fashion would allow the blood to flow. More tears, more heartache, more devastating thoughts about the future. Would he forever be in a wheelchair? What would this mean about his ability to grow, play sports, move through the world, fall in love, have a family, and so on?

And then resilience won again. The family, along with Bobby's cries for engagement in the world, began to integrate the wheelchair, finding ways to keep Bobby's world alive. Indeed, we even did some things that were probably not that safe. For instance, we lived next door to a churchhouse surrounded by a sidewalk. My older brother and I would push Bobby's wheelchair while running, timing how long it took each of us to go around the church – fastest time wins. Yes, we crashed a couple of times, and bruises and scrapes were part of it, but Bobby always asked for more…and my brother and I would run while pushing until we could run no more. And again, this is only part of the story. Seeing my brother in a wheelchair evoked many tears. He cried, too - a lot.

The wheelchair didn't work either.

As Bobby prepared for surgery, we prepared for what came next. The femoral ball had cracked and begun to fragment. Bobby would have to have his hip joint pinned back together. Following the surgery, Bobby was placed in a near full-body cast. The cast started just below his chest and ran down the length of his body to the end of his toes on the right side and to the top of his knee on the left. As with the brace, a metal bar was affixed to the cast, forcing his feet about three apart. Confined, trapped, and unable to move, Bobby spent eight weeks in his own plaster and metal prison.

The process of adjusting to change was at our door again. The challenges grew. Bobby had to be held over the toilet or a bedpan to use the bathroom (through a small hole in the plaster). Two of us would hold him by his armpits as he hovered over the toilet oddly. We bathed him on the kitchen table and let him lay there so he could eat with us – a new table centerpiece. He couldn't bend over as he did before to walk on all "fours." The cast prevented this possibility. Still, he flexed. He adapted. He began to using the walls, inching his way around the house, scooting along the floor using his one free leg and hands, rolling himself over and around – indeed moving across the living room like a turtle being moved by flipping on its back and then front and then back again. We laughed, he laughed…and we cried.

I tell this story not to compare it to Facioscapulohumeral muscular dystrophy (FSHD) or other muscular diseases but instead to point to something important

Dr. Kent Drescher shares in this book. He explores questions relevant to physical challenges: How do we live and engage under circumstances of being trapped by the body? How do you go on when faced with suffering, when fully challenged by life's unfairnesses, pain, and even cruelty? How do you build a meaningful life when FSHD is robbing your freedom to move?

In Acceptance and Commitment Therapy [ACT] for FSHD: A Psychologist's Personal Journey and Clinical Guide, Dr. Drescher offers answers to these questions. The book is comprehensive, describing the disorder, its short and long-term fallout, and an approach to living that supports the reader in resilience and meaningful living. He provides self-help exercises that help those suffering from FSHD to explore and discover new ways to relate to their psychological and emotional struggle with the disease, grounding the work in a practical understanding of what it means to have the diagnosis – given his own FSHD diagnosis. Dr. Drescher also provides personal stories of pain and resilience using ACT to locate the reader in an experience of authenticity concerning this progressive and debilitating disease. Finally, he addresses the experiences of caregivers and family members, assisting them to find meaningful living in the process as well. Acceptance and Commitment Therapy aims to discover how to vary and adapt behavior in an ever-changing and challenging circumstance through opening to emotional experience, becoming aware of the moment and all that it holds, and letting your values guide your choices as you engage life. Dr. Drescher brings this process into a vital and connected experience in the words he offers to those with FSHD.

I also want to add a personal note regarding my relationship with Kent (Dr. Drescher), which has now lasted more than 25 years. I have watched him use ACT to assist him through the many changes and difficulties he has experienced due to his own FSHD diagnosis. I am impressed by him again and again. This is not to say he hasn't had moments of deep pain and difficulty, but his resilience is fully evident. His work with ACT clearly helping him to live with purpose and connection. His friendships and family are at the top of the list concerning values well lived. I deeply appreciate Kent, and he is a model for me. I hope he can also be for you guided by what he presents here. My heartfelt thanks to Kent for his friendship and courage.

Yes, Bobby adapted to his physical challenges. He varied his behavior to meet the moment. But something else was happening, too. He and our family were meeting the moment with values-guided choices, finding love and meaning in the pain and joy of circumstances not selected. A journey not chosen but a journey engaged with presence and meaning. I hope you find the same through this book's personal and professional work – may it offer you a rich and vital approach to living with the challenge of FSHD.

Contents

Links to Exercises

PART TWO

PART THREE

Introduction

I have a chronic degenerative disease for which there is no cure, nor any effective treatment. I am 69 years old with a family and had a long wonderful career. My disease is called Facioscapulohumeral Dystrophy (FSHD), perhaps the most common form of Muscular Dystrophy (MD). My first symptoms of FSHD showed up 50+ years ago when I was in high school, though they did not diagnose me for another decade. These symptoms progressed slowly throughout the course of my life. My FSHD is significant, but not severe. I am still walking at least for short distances and can care for myself. However, not everyone is so fortunate.

I am a retired clinical psychologist who specialized in PTSD. For 27 years, I worked with military veterans as part of the National Center for PTSD and the VA Palo Alto Health Care System. I have a great family: my wife of 48 years, 2 sons, 2 daughters-in-law, and 5 wonderful grandchildren. I also have one sister. They have been my primary supports coping with this disease throughout the years. The emotional costs and burdens of this disease are high for those individuals afflicted, but also for their families.

I have been writing as a part of my professional career for 30 years. I've co-authored many professional articles, book chapters, and I have co-authored two books. But somehow, in all of that, I never imagined myself writing a book as a solo author, nor writing a book that is so personal.

A few particular events stand out when I think about the moments of my life that led to writing this book. For most of my life, living with FSHD was a solitary, even isolated experience. Having a progressive, untreatable, and incurable disease led me to avoid thinking about it, and avoid dealing with it as much as possible. Going to the doctor about it seemed pointless.

In the 33 years between my first diagnosis and 2016, I believe I only saw a doctor related to muscular dystrophy three times. The first was the initial diagnosis of MD. Six years later, when I developed a nagging, bothersome muscle pain, I saw a neurologist who clarified my diagnosis as FSHD. The third visit was another decade later when I started stumbling and falling more. For nearly all of that time, I just avoided dealing with the disease. I sort of felt like I had won the bad luck lottery, that had no cash prize. In all those years, I had met no one else with the disease. There was no one to tell me what to expect or how to cope.

Three experiences over the course of a year finally shook me out of that isolated and avoidant way of coping with my disease. Out of the blue, one day in April 2016, a book with an unusual title caught my attention. A young woman with the courage to write a book shared her own personal story and struggles with FSHD. Her book titled *The Graceful Art of Falling* offered me a glimpse into her personal journey, and I saw myself in her experience[1]. I no longer felt alone in the world.

The second event occurred in June 2016, when out of the blue, my wife and I got an invitation to attend an FSHD support group hosted at Stanford University. I now know that finding and registering myself on the FSHD society website led to that invitation. My wife and I attended the meeting. Sitting there in the conference room for two hours was a life-changing experience that I'll never forget. The people there were so very different, and yet in another way so very much the same. We were diverse, different ages, cultures, economic levels, physical abilities. Some were in wheelchairs, one person was on a ventilator, some used walkers, or a cane, or wore leg braces. Yet, looking around the room, there were people there who stood like me, moved their arms and hands like me. They had facial expressions that looked like mine. I felt a powerful sense of connectedness that I had never experienced before. I knew I had to find more ways of connecting with people that had this disease.

The third event that led to writing this book occurred a few months later. On the evening of November 30, 2016, I got a text message from my youngest son, Joel. It read:

> *"Hey dad, what form of muscular dystrophy do you have? Lauren and I are talking to her doctor about genetic testing, and she wants to know."*

When my son asked that question, I realized how little knowledge I had passed on to my sons about FSHD. I had known about the genetic test for the disease for over 15 years. I told myself that it seemed pointless to be tested as there was no treatment. Given our national healthcare system, for much of that time I was also afraid that having a genetic diagnosis might make getting health care more difficult for my family or even worse for my sons. I wanted them to make their own decisions about being tested. But I also wanted to continue to avoid having to think very much about the disease.

Years have now passed since those events, years filled with many non-FSHD life experiences, both painful and wonderful. I have come to terms with some of what avoiding the painful thoughts and emotions related to my FSHD has cost me.

I committed to write this book at start of 2020. I wrote most of it during the changed life experience we each had during the first year of the COVID-19 pandemic. My daily routine became getting up, sitting alone in the living room with my laptop, and writing. Sometimes it was productive and at other times not. The daily effort to apply ACT principles to my own FSHD while I wrote and lived life was both difficult and rewarding.

As a white male raised in middle class suburbia, I recognize I have lived a privileged existence my entire life. I have had access to unique educational and career opportunities that are not available to everyone in this society. I am also well aware that healthcare (and especially mental healthcare) access is expensive and even unavailable to many people in this society. FSHD affects people without regard to gender, race or culture, or economic status. Everyone affected should have access to the best resources for managing life with the disease, and to treatment when it becomes available.

My purpose in writing is to share the knowledge and expertise that life has privileged me to develop - with the entire FSHD community. There is a lack of literature that applies evidence-based mental health principles to the needs of the FSHD community. That's where my interest is, so that's what I wrote. This book will also apply to other forms of muscular dystrophy and likely to other neuromuscular and even neurological disorders. I'm going to leave it up to those patient groups to apply this book to those conditions.

Having struggled with FSHD for decades, I feel compelled to share what's been helpful to me with the FSHD community. I hope my time spent on this project will help those experiencing physical decline due to FSHD. I wrote this book as a self-help resource for those affected by FSHD and as a clinical guide for health providers.

I have learned many interesting things about FSHD as I have lived with it and in creating this guide. They classify it as a rare disease, though it may be one of the most common of the muscular dystrophies. A recent published prevalence rate suggests that FSHD affects 12 in 100,000 people worldwide[2]. This would equate to 40,000 people affected in the US, and a million worldwide.

It is natural with rare and incurable diseases that most research resources focus on finding a medical cure. The good news is that clinical trials underway have the potential to stop the progressive physical decline in patients. If successful, these treatments could become available within a few years. There's also a reason to hope that research will identify mechanisms for repairing and rebuilding muscle. This might bring about the possibility of a cure with a return to full healthy functioning. Unfortunately, this research focuses on the physical aspects of FSHD only.

There are mental health consequences stemming from FSHD and diseases like it. Mental health research and treatment can help people cope with symptoms of FSHD and other neuromuscular diseases. Living with FSHD and caring for those with it is costly, both emotionally and mentally, and resources are limited.

My years of professional and clinical work now seem relevant and beneficial for people like myself and my family who suffer from and care for those with FSHD. During later years of my career, I became interested in **Acceptance and Commitment Therapy (ACT) and received training**. ACT is a trans-diagnostic approach to treating many life challenges and problems. ACT helps people live purposefully and be more psychologically flexible, improving their quality of life.

When I first decided to receive training in ACT, it was because I saw its relevance for the patients that I was working with - military veterans with PTSD. I quickly learned however, that ACT views therapists and clients as peers. Both are human beings well acquainted with the struggles and pain of living. Therapists are taught to use the tools they are providing to clients within their own personal lives. I soon began to see that the tools I was sharing with clients, made an impact on me as well. They began to change the way I was living with my own FSHD and the relationship that I had with my body and my mind.

This book serves several purposes. I'll share my personal journey with the disease and its impact on my mental health and quality of life, as well as my family's. I'll summarize how FSHD affects mental health, as it can be challenging for laypeople to access and comprehend scientific studies. I'll describe research on how ACT can help FSHD patients cope with issues such as sadness, worry, pain, fatigue, and depression. I will share with you many practical exercises which have been helpful to me. They will hopefully be beneficial to you and your family members as well. Many neuromuscular and other chronic degenerative diseases cause similar physical and emotional struggles. This book may be relevant and useful for those individuals as well.

If you're struggling with the impact of FSHD, know that I'm writing this book for you because we share a similar journey. My deepest hope is that it will provide practical and useful tools that help you approach the life challenges of

FSHD more flexibly. I also hope it will help you move boldly toward the things that matter to you most. I hope your journey will succeed.

PART ONE

What is FSHD?

"People who have FSHD must cope with continuing, unrelenting and never-ending losses… no matter at which stage of life, the disease makes itself known, there's never after that any reprieve from continuing loss of physical ability…. FSHD affects the way you walk, the way you dress, the way you work, the way you wash, the way you sleep, the way you relate, the way you parent, the way you love, the way and where you live, the way people perceive you, interact with you, and treat you…. Each new day brings renewed awareness of the things you may not be able to do the next day."

- **Daniel Perez**, testimony before the US Congress

A few months ago, something happened that illustrates my recent experience with FSHD. My wife and I drove to the city of Benicia to visit my sister. I was already apprehensive as I wasn't sure I was feeling up to climbing the flight of concrete stairs to my sister's second-floor condo. Because of my FSHD, it has become a habit for me to think ahead of time about the potential obstacles I might encounter when I go somewhere.

After arriving, my sister came down to meet us and we went downtown together to get lunch. We drove to the waterfront area, parked. We had a delightful meal with a view of the water. After lunch, we went to get coffee and sit in the sun for a while to chat. It was a busy weekend day, so we had to park a distance from the coffee shop. It shouldn't have been a problem though, I had my folding electric wheelchair in the car. We got my chair out of the car and set it up on the sidewalk and I prepared to step into it to sit. Unexpectedly, my right foot caught on something, and my left knee buckled. Down I went.

I had fallen face-first onto the concrete. As a six foot two-inch tall man, almost all my falls are mighty. My right arm got trapped under my chest, and I was not strong enough to push myself up because of my FSHD. I lay on the sidewalk for a moment, gathering my wits. I finally rolled over to my side, grabbed my right knee with my right hand, and rocked myself into a sitting position. By this time, I noticed passersby standing close and asking if there was anything they could do to help. My wife and my sister listened to me to see what I needed. I still couldn't stand up. We positioned my chair and locked the wheels close to where I sat so I could grab the chair. I pulled myself up to my knees, and with some help, I pushed myself up high enough to engage my legs and get myself seated in the chair. We solved the physical challenge with just a scraped knee, a bruised elbow, and, as is often the case, a bruised ego.

Once the physical challenge was over, my mental challenge began. Emotions of embarrassment and anger emerged quickly after I fell. I always feel humiliated in these instances. Back in the wheelchair and settled in at a table outside the coffee shop, I sat alone with my thoughts while my wife and sister got coffee and cookies. My mind was racing, filled with worries, judgments, and new mental rules about how I should get in and out of the wheelchair. My thoughts were racing quickly: *"How did it happen THIS time?" "I'm such a useless, physical wreck," "What if I'd broken something?" "No one falls getting into a wheelchair!" "What happens when I can't get back into the chair?" "What if no one is around to help?"* Powerful emotions flooded into my awareness, including worry, fear, and hopelessness.

It hasn't always been like this. I was an athlete in my youth. I was full of energy and physical abilities - just like other young people. Until one minor event led to my diagnosis.

Thirty-seven years ago, I was a young, physically healthy man just beginning a new career after graduate school. Our first son was still an infant, and I had an amazing wife. I was sitting at my desk in my office in Boise, Idaho, typing away. I suddenly noticed I was only using seven fingers to type. The middle, ring, and pinky fingers on my right hand were curled in. Only my thumb and index finger were typing. I sat for a moment, baffled by what was happening; when had it started? I tried to extend my fingers and realized that those three fingers were not cooperating. They would no longer fully extend. It occurred to me I had just played racquetball with a friend a day or two before. While playing, I had stumbled and smashed my elbow into the sidewall of the court–it was still sore. I worried that maybe I had damaged something in my elbow that was affecting my fingers. I talked with my wife that evening, and we scheduled a doctor's appointment. After examining me, my family practice doctor told me he had good news and bad news. My elbow was undamaged. However, he referred me to see a neurologist.

In 1983, there was no confirmatory genetic test for FSHD. They made the diagnosis through clinical examination and a muscle biopsy. I remember the physical exam. The doctor carefully assessed the muscle strength throughout my body. I had to stand and walk while he observed my gait. I had to walk on my tiptoes and on my heels (I couldn't). A needle EMG was performed-where he would push the needle into a muscle and then ask me to move the muscle to measure the electrical activity. This happened multiple times in many places throughout my body - Ouch! He also did a nerve conduction test to rule out the presence of a nerve disorder such as multiple sclerosis (MS). He ordered a surgical muscle biopsy from my right deltoid muscle, which was sent out for analysis to the University of Utah. He asked about known family history (there was none). My parents flew out, and he physically examined and interviewed them as well.

Ultimately, the doctor told me I had a disease (myopathy) resulting from a genetic mutation that would likely slowly worsen. There wasn’t any treatment and it wouldn’t shorten my life. I don't remember him ever saying the words muscular dystrophy, and felt stunned when I walked out of his waiting room holding the medical billing form. I looked down to see that the diagnosis box checked was muscular dystrophy (MD).

The next day I went to the Boise State University library and looked up everything I could find on MD. As I reviewed the various MD forms in the library, FSHD seemed to me to be the most likely candidate. The formal diagnosis of FSHD occurred six years later, when I visited the USC medical center's neuromuscular clinic. More physical examinations and a second muscle biopsy (left biceps) produced the diagnosis.

The diagnostic process is simpler today. When physical indicators are present, a blood test sent to a special lab can confirm a diagnosis. However, some insurance providers hesitate to approve such testing due to cost. My son wanted to be tested as part of genetic counseling prior to having children. His health insurance would not approve the test unless there was a genetically confirmed diagnosis from a family member. As a result, I was finally tested and received genetic confirmation of my FSHD in 2017.

What I Learned

FSHD is a genetic disease that results in progressive weakening of the skeletal muscles, resulting in muscle tissue abnormalities and muscle tissue death. The condition is autosomal dominant. This means that each child born to a couple where one parent has the disease has a 50% likelihood of developing the disease. Nearly all individuals with the genetic defect develop symptoms of the disease (80%). Even those who develop no visible symptoms during their lifetime can transmit the disease to their children. As many as a third of cases have no apparent family history. However, some of this is likely due to never-tested and asymptomatic relatives.

FSHD (Facioscapulohumeral Muscular Dystrophy) is named for the muscle groups that are most typically and visibly affected first. Muscle weakness and loss are often first noticeable in facial muscles (Facio), back and shoulder muscles (scapula), and upper arm muscles (humeral). Over time, other areas such as the wrist and hand, lower abdominal, hip, and leg muscles may also become affected. It is common for FSHD to be asymmetrical. It often initially shows up only or is worse on one side of the body. It then eventually progresses to the other side, sometimes years later. Symptoms typically become visible during late adolescence or early adulthood, noticed by changes in athletic ability. While this is characteristic, some individuals don't present symptoms until their 50s or 60s. A small percentage (5%) exhibit severe symptoms before the age of five.

Muscle strength is integral to everything that we do. Weakness in certain muscle groups can dramatically affect an individual's ability to function in key domains. For example, weakness in facial muscles (mouth, lips, eyes) can cause painful chronic "dry eyes" because the lids of eyes don't fully close during sleep. Weakness in the mouth and lips can cause an inability to whistle, sip liquids through a straw, and impair speech clarity. The inability to smile can lead others to perceive the person with facial muscle weakening as overly serious, sad, or even hostile.

Arm and shoulder weakness can lead to an inability to raise one's hands above shoulder height. This can make it hard to push up from the floor or bed. It can also make self-care activities (brushing teeth, washing hair, getting dressed) difficult. Hand weakness can limit your ability to do everyday things like eating, working, and enjoying hobbies. Standing or walking may be harder with weak abdominal muscles. This can cause postural and skeletal issues, such as lordosis. To the degree that the diaphragm becomes involved, it can compromise breathing.

Leg weakness can lead to foot drop and cause people to stumble and fall. It can make climbing stairs difficult or impossible. Weak upper and lower body muscles can make standing up, transferring, or using the toilet difficult. It can be difficult for a person to perform important activities such as dressing, grooming, and eating because of this. Sometimes a paid care provider becomes necessary to help with important daily activities. Approximately 20% of individuals with FSHD will need a wheelchair full time by the age of 50. Some will require full-time attendants or caregivers to help them take care of the basic activities of daily living.

When I first learned that I had a rare disease, I felt alone with my experience. No one in my family understood, and for nearly 40 years I never met anyone else with the disease. Most of my doctors through the years were unfamiliar with the disease or its impact. The U.S. designates diseases that occur in fewer than 1 in 5000 (or 20 per 100,000) individuals as rare. Until just recently, the estimated prevalence rate of FSHD was 5 per 100,000 people.[1] The most recently published study increased that rate to 12 per 100,000.[2]

Consider what this means. The total population of the US in 2020 is about 331 million people. An FSHD prevalence rate of five per 100,000 means that throughout the US there are only 16,500 people with the disease. If the rate is 12 per 100,000, it means that there are approximately 40,000 people with FSHD in the US. At the lower prevalence rate, you could seat every person with FSHD in Madison Square Garden to watch a Knicks' game, and it wouldn't be a sellout. There would be 4,000 empty seats. If the prevalence is the higher rate, you could send everyone in the U.S. with FSHD to see the Yankees play at Yankee Stadium, and there would be 16,000 empty seats.

Whether at the Knicks or Yankees, FSHD would make it difficult for many attendees to climb the stadium stairs to find their seat. Some of us with FSHD could not take part at all without a wheelchair.

An imaginary visit to a stadium filled with FSHD patients illustrates how rare the disease is. The reality is that those few people are scattered across an immense country. We rarely encounter others with the disease. Because of this, people with FSHD often suffer in isolation. They may never or only infrequently meet someone that understands their struggle.

The Status and Need for Research

Having a rare disease often means knowing a cure may not come quickly.

Treatment and a cure require research. We funnel most federal research dollars toward other diseases that effect more people. It's hard to attract public and private funding for research when a condition is rare. Companies often consider the research and development costs too large to be cost effective. Spending a massive amount of money to develop a medication sold to a tiny number of people will never be profitable for a pharmaceutical company. The small number of affected individuals can also make it hard to recruit sufficient clinical trial subjects and require novel research designs specific to small populations.

It is also hard to find sufficient expertise within the government agencies that oversee and review research applications and that allow the marketing of medications and other products. The federal government has taken steps to increase speed up and support research and product development for rare diseases.[3] However, with FSHD, the scientific progress toward developing a cure or any effective treatment has been slow.

There is hope. The physical, emotional, and financial costs of rare diseases motivate patients, family, and friends to advocate for research and treatment development. In recent years, advocacy groups have partnered with academic researchers, pharmaceutical companies, and the government to hasten scientific progress. The FSHD Society and the Muscular Dystrophy Association are important advocacy groups in the U.S. that facilitate research partnerships and expedite the development and approval of treatments.

The Problem of Isolation

Those with FSHD rarely meet or interact with anyone else that has the disease, except possibly family members. For the 30% of patients who present with no known family history, it's challenging to know what to expect for the future. It’s also difficult to find support from people that understand your experience. Few doctors and health providers know about the disease unless they work in a specialized neuromuscular clinic that is connected to ongoing research. As noted, from my initial diagnosis in 1983 to 2016, I never met another person with the disease.

Finally, I heard about and attended a patient meeting sponsored by the FSHD Society at Stanford University. This was when I first met other people like me. That first meeting was a powerful experience. It was overwhelming seeing the varying range of symptom severity in the room. I could see similarities in facial expression, body posture and movement. I saw teenagers and old people. A family member drove one young woman nearly 400 miles just to be there. We differed in age, gender, ethnicity, and economic status, but we shared one thing in common.

I experienced a potent mix of emotions at that meeting. I felt a powerful sense of connection. I felt fear about what might lie ahead–seeing several people in wheelchairs, one using a ventilator to breathe. But I also experienced a sense of relief that here were people present that might understand me. Seeing that my symptoms weren’t as severe as some present left me feeling both fortunate and a little guilty. Yet, somehow inexplicably, I experienced the sense that “These were my people,” I was home at last. Even now, as I write about this experience, I sit with tears in my eyes, feeling those emotions again through my memory of the event.

Living with a disease with no cure or treatment that progresses at an uncertain rate can elicit various emotions. You may feel frightened, confused, frustrated, and anxious all simultaneously. You may have questions like, “Why is this happening?” and may feel mistrust or feelings of blame. I have sometimes felt hopeless about my future. One can turn bitter and angry, withdraw from loved ones and activities, and even hate or punish their own body. In short, there are many ways in which FSHD can affect one’s mental health and quality of life.

What is known about FSHD and Mental Health?

FSHD has an enormous physical impact on an individual’s ability to live an active life and take care of their own financial and personal needs. So, it’s not surprising that virtually all the research dollars mentioned above focus primarily on basic research toward a cure. The initial goals are to understand the mechanisms that produce symptoms, and to develop strategies to stop the disease's progress. The ultimate aim is to return strength to muscles and improve functional ability. In other words, the primary investment is in finding a cure. Given this focus, it's also not surprising that very few studies have examined the emotional and mental health impact of living with FSHD right now, today. However, understanding how the disease affects emotional and mental health is important as well. These things, too, are part of the disease’s impact and process.

The rest of this chapter reviews research on the impact of FSHD and similar neuromuscular conditions on mental health and quality of life. Often, my personal experience with FSHD matches these findings.

Thoughts and emotions and FSHD

One of the FSHD Society founders quoted earlier said: "*People who have FSHD must cope with continuing, unrelenting and never-ending losses…*" Sadness and grief following a loss are potent emotions. Those feelings can become overwhelming over a lifetime, experiencing the progressive loss of physical function that results from FSHD. I mentioned earlier the event that led to my diagnosis with FSHD. The fingers of my right hand's extensor muscles failed, which I first noticed affecting my ability to type. It was months later when I realized the symptom had dramatically affected something else that was even more important to me–my ability to play music.

During junior high and high school, I took music lessons for classical guitar. It was something that I loved. It was important to me. At the end of high school, I saved all my earned money from working during the summer of my senior year to have a handmade classical guitar custom built. I picked out the wood for its construction and went every week to the guitar maker's shop to observe his progress as the instrument took shape. It was a beautiful instrument, and I loved playing it. However, the unimpaired use of the fingers of one's right hand is necessary to play classical guitar. Without warning, this disease had taken away one of the most important activities of my early adulthood. My feelings of sadness and loss were powerful. When a friend from college (who also played guitar) traveled with his wife to our home for a visit, I asked him to take my guitar home with him. I couldn't emotionally bear to have it in my house anymore. The guitar was a daily reminder of my loss. It was five years before I could go to his house and ask to take it back.

Research data shows that FSHD often proceeds with a slow, progressive weakening in muscle groups throughout the body. However, patients often describe their experience not as a steady decline but more like stepping down a staircase. They describe periods that seem like a plateau where they notice no physical decline. This is followed by a rapid change that results in a noticeable loss (or change) in function.

In high school, I took part in track and field and tennis. In college, I played on the men's volleyball team. Those activities continued beyond my college years. Sometime around my diagnosis, without awareness of the reasons, my tennis playing transitioned to racquetball (smaller lighter racket, smaller court), and running transitioned to bicycling. I rode a bicycle regularly for much of the next 25 years. But even with cycling, there were changes across the years. I transitioned from a road bike (down-turned racing handlebars) to a folding road bike with no top bar to step over. I later converted to more upright handlebars after I experienced wrist pain and arm strength loss. Even later, getting on and off the bicycle seat became increasingly risky, and I fell getting on the bike and broke a finger. I then switched to a cruiser-style bike with "flat-foot technology" - that is - where I could place both feet flat on the ground while still sitting on the seat. This eased getting on and off the bike and made me more stable when starting and stopping the bicycle at traffic signals.

Some think that what happens with FSHD is that a long period of slight decline reaches a threshold where a muscle can no longer perform a significant task. When that comes into the person's awareness, a work around becomes necessary. Suppose that function is important to life quality. In that case, the person might notice it in their awareness as a sudden significant change. Physical activities and exercise are one way I have observed the changes in my body over the years. When changes in physical functioning start requiring adaptive devices, we can think them of as transitions. Transitions can occur suddenly, or they may be more subtle and experienced over a longer time. The emotional stress of repeated transitions may put individuals at risk for mood problems, such as anxiety or depression.

Emotional and social impact of living with FSHD

Quantitative studies of large representative samples of patients provide important information about how often and how intensely certain FSHD symptoms occur. However, qualitative research is also useful. Qualitative studies start with small groups of patients to identify important themes for later research.

Qualitative studies do broad open-ended interviews of a few people. They carefully analyze interview transcripts for unique themes that emerge. Johnson and colleagues performed one such study with 20 adult FSHD patients. They sought to identify unique perspectives on which symptoms have the most significant effect on their lives.[4] This study's findings ultimately provided the framework for developing the survey used in an important quantitative study–the PRISM-FSHD Study.[4] The Johnson study took 1,375 quotes from the interviews and from those identified 251 unique symptoms that were grouped into 14 themes. Several of those themes are mental health challenges. These include social role limitations, emotional distress, social role satisfaction, and impaired body image.

The PRISM-FSHD study analyzed patient perceptions of hundreds of FSHD symptoms to determ*i*ne the most impactful ones.. The study surveyed 328 individuals with FSHD about the frequency and impact of these symptoms. Participants were a nationally representative sample drawn from the FSHD patient registry. Individuals from 47 different states in the U.S. took part.

As one would expect in a study of FSHD, by far most of the symptoms are physical. The symptoms reported with the highest frequency and intensity are key physical indicators of FSHD. For example, 96.9% of participants reported problems with shoulders or arms. Older participants reported greater impact because of the progressive nature of the disease. 93.6% of respondents reported walking or mobility limitations, and this impact was greater with older age. Chronic fatigue (93.8%) and pain (87.7%) were also highly prevalent.

When I first read the study, I realized how many of the physical symptoms were things I had experienced during my disease. I also found that I identified with the emotional and social struggles examined in the study. Let’s turn to some of what the PRISM study says about these issues.

Participants reported frequent and intense mental, emotional, and social issues. I have struggled, perhaps like you, with a changed body image. Indeed, I have felt uncomfortable when I notice people staring at the way I stand or walk. It bothers me when random people ask me intrusive personal questions that they wouldn't ask someone else. I often feel uncomfortable in social situations when I feel my FSHD symptoms make me the focus of attention, or I feel like an object of sympathy or pity.

The PRISM study found that experiences like these are common. Nine out of ten FSHD patients report changes in their body image. Three out of four report emotional distress from FSHD, difficulty performing in social situations, and reduced satisfaction in social situations.

The PRISM study also shows that the mental, emotional, and social impact of FSHD is lower in FSHD patients who report little or no chronic pain. Similarly, the impact of emotional issues and changed body image was significantly lower for those with minimal or no pain. The PRISM study clearly shows how important it is to address emotional and social problems for individuals with FSHD, (especially for those with high pain levels) given the high frequency in which they occur.[5]

Physical pain and FSHD–the intersection of pain and mental health

I feel fortunate that so far - my physical pain has only been occasional as opposed to chronic. I have only experienced severe pain two or three times during my life, where it lasted for days and was intense. Luckily for me, in those instances, physical therapy and massage over a period of a few weeks reduced my pain to a manageable level. I know, however, that this is not the experience of many people with FSHD who deal with excruciating, unrelenting pain daily.

Chronic physical pain is not unique to FSHD. The National Center for Healthcare Statistics (NCHS) reports that 25% of the US population has recurrent pain. Ten percent report pain lasting more than a year, and 40% of those folks report that pain moderately or severely impairs their functioning.[6] Many in our society have pain from accidental injury. Pain also results from many diseases. Because of this, much research exists on the impact and treatment of chronic pain.

For many years, research studies and review articles have documented the co-occurrence of chronic pain and depression. Results of the studies suggest that this association works in both directions. Having chronic pain increases the likelihood that a person will develop depression. However, the opposite is also true. Having depression also increases the likelihood that a person will experience chronic pain. A recent study showed that chronic pain sufferers have an increased risk of developing depression.[7]. Experiencing pain in multiple locations in the body and having more severe pain resulted in increased depression.

Several studies have examined the emotional and mental impact of chronic pain in neuromuscular patients, including muscular dystrophy. Pain and fatigue are associated with physical impairment and depression in muscular dystrophy patients, including FSHD, according to one study.[8] The study suggested under-utilization of psychological pain interventions within this group and recommended their increased use. Several other FSHD patient studies also show that physical pain results in significant worsening in quality of life–including mental and emotional life quality.[9-11]

Chronic fatigue and FSHD–the intersection of fatigue and mental health

Fatigue is a daily challenge for me. The level of exertion required to do the simplest things is still striking to me, when I think of how things used to be before my symptoms worsened. Climbing a flight of stairs can now leave me exhausted. Going for a short stroll with my family has me rapidly looking for a bench to sit down as soon as we get started. Everyday chores, such as emptying the dishwasher and lifting plates onto the lowest shelf of the cupboard, are tiring. I'm sure I'm not alone in those experiences.

In the PRISM-FSHD study, fatigue was reported by 94% of FSHD patients.[5] Individuals with extreme fatigue also reported less motivation, physical activity, and worse mental health.[12]

The second study was a longitudinal study that contacted participants over an 18-month timeframe. Low muscle strength, physical inactivity, sleep problems, and pain all contributed to higher fatigue in the FSHD group. Lack of physical activity and high fatigue both led to more impairment in important life activities.[13] Considering this finding, Voet and colleagues conducted a randomized controlled trial to test whether regular aerobic exercise or cognitive-behavioral therapy would decrease fatigue among FSHD patients.[14] The study found that both interventions resulted in a significantly reduced fatigue. These improvements continued for several months after the completion of the intervention. Physical activity increased with both interventions, but only cognitive-behavioral therapy improved sleep and social engagement.

Depression and Anxiety in FSHD

Only one study has examined the prevalence of mental health disorders in people with muscular dystrophy, including FSHD. Kalkman and colleagues performed diagnostic interviews with 217 patients with neuromuscular disorders. (65 had FSHD).[15] Researchers consider a careful clinical interview to be the most accurate in diagnosing mental health disorders. This study found that 31% of the FSHD patients had a history of a psychological disorder during their lifetime, and 12% within the previous month. The lifetime rate for depression was 17%, with 5% of those occurring in the past month. The lifetime rate for anxiety problems was 15%, with 4% occurring in the past 30 days. None of these rates are significantly higher than rates found within the general U.S. population. This suggests that FSHD may not convey a much higher risk of developing mental health disorders than the general population. That being said, at least one small study found that individuals with FSHD scored significantly higher on screening measures for depression than did control subjects in the general population.[16] It's also important to note that one does not have to meet the full diagnostic criteria for depression for depressed mood symptoms to affect one’s life and functioning. Based on my experience, I can say that the challenges associated with FSHD have contributed to my experience of depression, but not always as the primary source. People with FSHD also have to contend with all the other everyday life struggles that make depression and anxiety so common in our society.

The role of emotion – loss, fear, sadness

Emotions can play a prominent role in the lives of individuals with FSHD, just as they do in those who do not have the disease. Daniel Perez’s words quoted at the beginning of this chapter mention enduring losses throughout the disease's life. Enduring awareness of loss and sadness has been a fixture for me during my disease progression. The awareness of losses is constant. However, the emotional experience of sadness arises at specific times when I am particularly reminded of these losses.

As I’ve mentioned, intense emotions of anger, humiliation, and embarrassment can intensely flood my awareness when I fall or feel on display. Worry is a cognitive phenomenon associated with recurrent thoughts, accompanied by emotions such as fear, anxiety, and hopelessness. Worry and anxiety about the future is something that I experience frequently and that I've heard recounted by many other individuals with FSHD. Little research focuses directly on the emotional experience of individuals with FSHD. However, one study found indications that individuals with neuromuscular disorders frequently share their emotional experiences in online forums.[17] They completed a thematic analysis of the first five months of the content of the posts. Many posts expressed emotions about their illness, anxiety about the future, and feeling unsupported by healthcare systems.

Nearly every day, individuals with FSHD post questions, share worries and fears, and receive encouragement and support from a Facebook community called Living with FSHD. I’ve experienced many of these concerns. If you have these experiences, know that you are not alone.

The role of cognitions and beliefs about symptoms

Though research dollars are scarce, research into how thoughts and emotions affect the lives of those with FSHD is still important. It is important for a better understanding of the role the mind plays in the person’s experience of FSHD. Identifying ways for individuals to live full and meaningful lives with FSHD is crucial, given that there still is no effective treatment or cure.

As mentioned, research shows cognitive-behavioral therapy (CBT) can help address issues of pain and fatigue with FSHD. According to CBT theory, one's thoughts, beliefs, and attitudes affect the development and treatment of mental health issues. With many chronic diseases, research shows that specific thoughts and beliefs related to an individual's condition can play either a helpful or harmful role in disease outcomes.[18] These "illness perceptions" impact mood and mental health. They also affect a person's adherence to treatment recommendations, and with some diseases, even influence mortality.[19] Research is underway with many diseases to understand how to change illness perceptions to improve patient outcomes.

Christopher Graham and colleagues have conducted several studies related to illness perceptions in individuals with neuromuscular conditions, including FSHD.[20-21]

Dr. Graham was interested in learning about how thoughts and beliefs impact muscular dystrophy. He was also interested in how quality of life, mood and ability to function relate to their thoughts and beliefs about their disease. He found that study participants grouped into three clusters. One group contained individuals with the worst functional impairment and disease severity. These individuals had the lowest quality of life and most problems with depression.

Group two and three had similar levels of functional impairment, but differed widely in their experience of the disease. Group two had a better quality of life and better mood than either of the other two groups. Those in this group had more helpful illness perceptions than Group three, and more confidence that they understood their disease and its impact. They recognized MD was lifelong, and slowly worsening. They more accurately distinguished the symptoms connected with their disease from other unrelated symptoms. This group also had less emotional volatility and more acceptance of their disease than Group three. Despite having the same disease severity and functional impairment as group three, group two had a better quality of life and fewer mood problems. Researchers recommend providing disease education for MD patients based on its success for other illnesses.

The research identified important parts of a mental health intervention that boost outcomes for those with muscle disease, including FSHD. It noted that the education should encourage acceptance of disease progression as a core element. The second aim is to lower emotional volatility around how the person understands and experiences their illness. However, they emphasize the need to recognize the natural fear and frustration of having an incurable progressive illness. They propose that an intervention focus on identifying and reconnecting with important values. This helps reduce the power of emotions, such as fear, sadness, and loss. They should pair these values with actions that actualize the values in the service of a life that can be meaningful, even in the face of mobility challenges.

Dr. Graham's team did another study on how illness perceptions, coping, and optimism affect the mood and quality of life of people with neuromuscular disease.20 It included disease factors such as weakness, pain, fatigue, and other things that impact lifestyles like independence, activities, social connectedness, and emotional health. Coping and optimism made a small impact on the quality of life. Helpful illness perceptions also predicted lower anxiety and depression. Here, coping and optimism played a larger role in mood than in quality of life. However, illness perceptions still made the greatest impact on mood.

Dr. Graham's team also researched the role that psychological flexibility plays in the lives of people with neuromuscular disease.[22] The study surveyed 136 respondents (28 with FSHD) at two time points 4-months apart. Its focus was to see whether psychological flexibility improved mood and life satisfaction more than other factors (disease severity and illness perception). Results showed that psychological flexibility predicted improved both life satisfaction and mood.

These studies give me hope that I can improve my quality of my life by learning more about FSHD and by becoming more psychologically flexible.

The proposed role for Acceptance and Commitment Therapy (ACT) for FSHD

These studies suggest that psychological intervention could be useful for individuals with neuromuscular diseases. Dr. Graham and colleagues have established the groundwork for exploring the potential usefulness of Acceptance Commitment Therapy (ACT), a third-wave behavioral therapy. His team reviewed the existing literature to examine findings from previous studies. They wrote one review about quality of life among neuromuscular patients.[23] They wrote a second paper reviewing research on whether ACT is helpful in other chronic degenerative disease conditions.[24] Their review found solid evidence supporting an association between quality of life, mood, and illness perceptions. Each of these factors are targets for psychological interventions. They also found that pain and fatigue reduced quality of life. There is promising support for the use of psychological interventions related to both pain and fatigue.

The second review paper examining the impact of ACT found modest support for using this approach with other chronic conditions.[24] Though existing studies found consistent support for the helpfulness of ACT with these conditions, there were few randomized control trials (RCT), and the sample sizes for those trials were quite small. This means that there is still much research to be done.

Dr. Graham and colleagues have now completed two preliminary studies of ACT's usefulness with neurological and neuromuscular diseases.[24-25] One pilot study explored how eight patients with neurological disorders responded to ACT.[25] The therapy intended to reduce disease symptom interference with life functioning and to improve mood. Researchers hoped that increased psychological flexibility would produce clinical improvements. Five of the eight patients reduced symptom interference with life, and four showed improved mood. None of the participants showed a worsening of mood or symptom interference. Psychological flexibility varied within the sample.

The second study administered ACT in a self-help format to eight people with neuromuscular diseases hoping to improve quality-of-life and mood.[26] Using a self-help format is important because it broadens the intervention's potential impact. Individuals with mobility difficulties often find it challenging to seek psychological treatment in a clinic. If studies show self-help psychological interventions help people with muscle diseases, more people can access and benefit from them. The self-help intervention had seven participants. It contained three ACT modules that were completed over a three-week timeframe. Each client received a 15 to a 20-minute phone call at the end of each week of the intervention. Four of the seven participants improved on measures of quality of life, anxiety, and depression. Psychological flexibility did not show consistent improvement. No participants showed worsening on any of the outcomes, and feedback about the intervention was positive.

A randomized clinical trial (RCT) is the gold standard for establishing whether a treatment works. Based on the work described above, Michael Rose and colleagues (including Dr. Graham) conducted an RCT of a self-help ACT intervention for individuals with neuromuscular diseases, including FSHD.[27] The trial began in 2016 and completed recruitment and interventions in 2019. The intervention involved four modules comprising ACT information and exercises. Each participant in both the control and intervention conditions also received standard medical care. Participants in the ACT intervention group also had a phone conversation with a clinician providing guidance.

Initial results of the RCT show mood and Quality of Life improvements in 139 patients with neuromuscular disease (50% FSHD) through self-help Acceptance and Commitment Therapy (ACT). The study randomized participants into two groups. One group received standard medical care (SMC), and the other received ACT plus SMC. Researchers saw increasing improvements in QOL at 3, 6, and 9 weeks following treatment in the ACT group. The ACT treatment involved completing four weekly written / audio exercises at home (45-90 minutes to complete). Each participant also received a weekly therapist phone call (15-30 mins) to review the weekly materials and answer questions.

The ACT group saw medium to large effect improvements on the primary outcome measure — the *Individualised Neuromuscular Quality of Life Questionnaire* (INQol). It measures the impact of muscle disease on patient activity, independence, social functioning, emotional functioning, and body image. There were also improvements in secondary outcomes, including anxiety, depression, psychological flexibility, pain, fatigue, and social functioning. This study is only the third clinical trial to show positive benefit from psychological intervention for muscle disease, and the first that targets outcomes broader than physical activity/fatigue.

For me, Dr. Graham's interest in ACT to improve the lives of people with FSHD and other muscle diseases is important. His team's work has spurred me to examine how I can use these principles to improve my life. I am grateful because it has helped me, and I believe it can help you as well.

PART TWO

Exploring Acceptance and Commitment Therapy (ACT) as an Intervention

suf·fer·ing "the state of undergoing pain, distress, or hardship."

— Google Dictionary

"To live is to suffer, to survive is to find some meaning in the suffering."

— Friedrich Nietzsche

Universal human suffering

The experience of pain and suffering is seemingly universal within the human species. In this book we will attempt to distinguish between pain and suffering. Pain is both physical and emotional, and is felt as sensation within the body. Suffering tends to be a mental experience. We live in a world where disease, poverty, hatred, oppression, injustice, discrimination, and violence are common and create significant pain and suffering. Additionally, even when human beings manage to escape those challenges and possess all the things traditionally associated with "success" (items such as financial security, a loving, caring family, even fame and fortune), they can still be miserable. U.S. statistics indicate that the lifetime prevalence of psychological disorders is now approaching 50% of the US population. This does not include additional emotional distress associated with relationships, families, jobs, and specific developmental periods such as adolescence and aging.[1]

Dr. Steven C. Hayes, the primary developer of Acceptance and Commitment Therapy (ACT) summarizes the prevalence of psychological suffering this way:

> *"If we add up all those humans who are or have been depressed, addicted, anxious, angry, self-destructive, alienated, worried, compulsive, workaholic, insecure, painfully shy, divorced, avoidant of intimacy, and stressed, we are compelled to reach a startling conclusion, namely, that psychological suffering is a basic characteristic of human life."* **2(p4)**

As human beings, individuals with FSHD experience suffering associated with all of the things mentioned above at rates similar to all other human race members. However, individuals with FSHD also carry the additional burdens and suffering associated with having the disease. This suffering is both physical and mental. Severe and sometimes enduring physical pain results from injuries received from falls, or from the skeletal misalignment associated with the weakening of core muscles, or from partial and even full joint dislocations related to the deterioration of surrounding muscle tissue. Pain can also result from the overuse of small muscles attempting to support weakened larger muscles.

Mental suffering for individuals with FSHD arises in part from the knowledge they are experiencing (and in some cases have passed on to their children) a progressive, degenerative, and sometimes debilitating disease for which there is currently no treatment or cure. Individuals with FSHD live with the daily awareness of the physical abilities the disease has taken away in the past, often worry and ruminate about what the next loss will be, and wonder how and whether they will be able to cope. These physical functioning losses can impact the individual's ability to find and hold a job, build and keep relationships, and care for themselves and others without assistance. It's not surprising that individuals with FSHD might experience depressed mood, anxiety, intense anger and frustration, a reduced sense of life satisfaction, and ultimately an impaired quality of life.

Stigma, bias, disability, and intersectionality in the context of human suffering.

Before we turn to look at how ACT understands suffering, I'd like to turn attention to defining some terminology that is relevant to our discussion of human suffering. We will make occasional reference to these throughout the remainder of the book. The first term is **stigma**. A dictionary definition of the word is: "*a set of negative and unfair beliefs that a society or group of people have about something.*" Stigma takes many forms in our world today. People, places, and activities can all have stigma attached by society. Groups of people who different from the majority on the basis of race, ethnicity, gender, sexual orientation, or disability often have stigma directed toward them. Activities such as seeking mental healthcare also are stigmatized. Stigma may motivate people to withdraw and avoid situations that would bring attention to their source of stigma.

Bias is defined as "*a personal and sometimes unreasoned judgment.*"Another word for the term bias is prejudice. Bias can be either explicit, or implicit. Examples of explicit bias would be active discrimination or hate speech. Implicit bias is prejudice that is measurable but exists outside of the individual's conscious awareness. For example, the majority of our society shares an implicit bias that unconsciously associates science and technology with male gender. So, from a very young age young girls are not encouraged by people around them to pursue those careers. Many people hold implicit biases related to disability and often these unrecognized beliefs impact opportunities and resources available to the disabled individual.

The APA dictionary defines **disability** as "*a lasting physical or mental impairment that significantly interferes with an individual's ability to function in one or more central life activities, such as self-care, ambulation, communication, social interaction, sexual expression, or employment.*" Not everyone with FSHD is or will become disabled. However for many, FSHD will at some point in their life create difficulty functioning in one or more of those areas listed.

The final term that I want to mention is **intersectionality.** Kimberlé Crenshaw coined this term to describe how discrimination against different multiple facets of a person's identity can overlap and impact their lives. Race, ethnicity, gender, disability, and sexual orientation all interact to affect a person's lived experience and contribute to unequal outcomes in ways that cannot be attributed to one dimension alone. As my disease symptoms have worsened and my mobility declined, I have begun to struggle with the subtle and obvious obstacles that society creates for those with mobility challenges. I experience those things as a financially secure White male. Intersectionality means that that those challenges are multiplied for individuals that experience additional stigma, bias, and discrimination due to other aspects of their identity. Intersectionality increases suffering for those individuals, and makes life more difficult.

How does ACT understand human suffering?

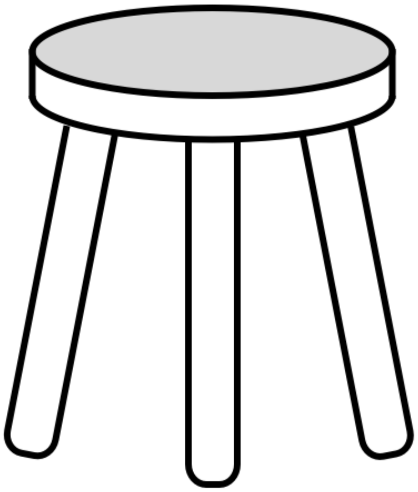
Historically, philosophers and religious leaders have addressed the challenge posed by the universality of human suffering. For the most part, these approaches focused on the "Why" of human pain and suffering. Theodicy is a term introduced by the philosopher G.W. Leibniz. His question went something like this: If God is all good, and God is all-powerful, why does human suffering exist? For if God were all good, God would prevent suffering, and if God were all-powerful, God could prevent suffering. While philosophical and religious questions about why suffering exists continue to be discussed, the modern age has brought about the scientific focus on the physical and mental causes of pain and suffering.

Since the dawn of the scientific era, and particularly in the last 100 years, human pain and suffering have increasingly been "medicalized." Physicians have viewed pain and suffering as symptoms of disease or injury and focused on developing treatments to eliminate them. This has undoubtedly resulted in remarkable breakthroughs in disease care. However, in recent years the intense focus on eliminating pain through medications has also produced horrific unintended consequences.

ACT also has a perspective on human suffering, but rather than address the metaphysical origins of human suffering or medicalize it, ACT seeks to distinguish between pain and suffering. ACT simply acknowledges the universality of physical, emotional, and spiritual pain among humans. From an ACT perspective, suffering comes about when human beings engage in an intense internal struggle with pain of all sorts. From this perspective, struggling with pain (i.e., unhelpful and unworkable attempts to avoid and control one's bodily sensations, thoughts, emotions, and memories) not only fails to relieve pain, but may actually intensify and prolong the experience. Each present moment spent avoiding pain and struggling internally with difficult thoughts and emotions steals away the opportunity for experiencing meaning and satisfaction derived from living one's values.

ACT for FSHD

ACT seeks to increase human flourishing even in the presence of mental and physical pain, by increasing what it calls "psychological flexibility." Psychological flexibility is the "*ability to respond to life in a workable way, a way that enables us to live vital, meaningful lives.*"[3(p9)] To accomplish this new way of living, ACT engages individuals in three distinct therapeutic practices. These practices can be thought of as being like the legs of a three-legged stool. As with a stool, each leg shares equal importance in supporting psychological flexibility.



The Three-legged stool of ACT

The first of these practices, **Opening Up**, involves increasing an individual's willingness to experience (Acceptance) everything that life brings without engaging in unhelpful avoidance or attempts to control one's internal emotional experience. It also encourages holding less tightly (Defusion) to various thoughts that would discourage one from engaging in values-directed actions.

The second of these practices, **Awareness,** involves developing the practice of experiencing the present moment using mindfulness (Present Moment). Jon Kabat-Zinn, a noted expert in the field who taught at University of Massachusetts Medical School, defines mindfulness as *"paying attention in a particular way: on purpose, in the present moment, and non-judgmentally."*[4] Awareness also involves strengthening one's ability to notice their inner experience, as well as to notice the enduring "observer" part of the self. It is that observer that can step back from experience and choose one's actions in each moment (Self as Context).

Finally, the third of these practices is **Doing What Matters**. This involves a continuing search for and pursuit of one's most important values (Values), along with a moment-by-moment commitment to take actions on those values (Committed Action).

The Promise and Challenge of human language

As human beings, we live our lives within two worlds. The first is the outer world. This is the physical world where we move about and work. It is where we have a variety of memorable and intense experiences, including relationships with other people. The second is the inner world. This is the world of emotions, memories, sensations, and thoughts. This world is ours alone, except to the degree we use language to describe that inner experience to others. Emotions are an exception. Emotions are a non-verbal and sometimes unintentional means of communication with others. Our emotions tend to leak out through facial and bodily expressions, postures, and vocal tones, even without our permission.

The human mind is highly adapted to creative problem-solving in the physical world. It brought our species to the top of the food chain. It allowed us to go from small groups of cave-dwelling hunter-gatherers to a large society of highly technologically sophisticated individuals. The human mind invents impressive gadgets and technological devices, allowing for the communication and storage of massive quantities of information and history. This advancement and change in our physical world has occurred over a relatively tiny timeline of perhaps 12,000 years.

The human mind is a highly flexible, creative tool for solving problems in the physical world. Human language is at the core of that ability. Language allows for efficient communication (through speech and writing) and storage (through memory and writing). Through languaging processes, our brains mentally envision new ideas. They can imagine things never seen in the physical world. They then envision the necessary steps to create them and then analyze and evaluate their usefulness – all without ever actually seeing or touching them in the physical world. It is this capacity that drives human flourishing in the physical world.

The foundation of that creative capacity is the mind’s ability to mentally form associations or relations between things that don't actually exist. Language itself exemplifies this ability. Look for a moment at these three discrete shapes below.

Y A M

By themselves, these are simply scribbled shapes that we choose to call letters, each having a different form. These letters can be arranged into words, each of which can have a different meaning. Place these letters together in one order - YAM - and we find a meaning we likely learned in school or in the kitchen. We are now mentally picturing a root vegetable similar to a sweet potato.

If we flip those letters around the word becomes - MAY - and suddenly we're imagining a calendar showing the fifth month of the year. Rearrange the letters yet again and we have – AMY - the name of a person, perhaps the name of a friend. Each of these associations was learned. Notice how each rearrangement of the same letters leads you to imagine something different.

Let's look at this further. Depending on how the letters are arranged and the context in which they are used, we can connect them to memories and emotions and actual relationships in the physical world. AMY might be someone I met or married. YAM might be a favorite food tied to pleasant memories of family gatherings across a lifetime. MAY might be the month I had an amazing adventure. Rearranging, and sorting letters and words creates millions of relations and other associations. This is the work of the mind. This work is non-stop. It happens moment after moment, day after day, and mostly outside of our conscious awareness.

If I were reading this instead of writing it, I imagine my mind might be saying something like, "*Where is he going with this? Why is he telling me this?*" *Why does any of this matter? What can it possibly have to do with FSHD?*" If that is what your mind is saying to you - I'm going to ask you to try to be patient for a few moments. It's going to take me a bit of time to explain. But the things that our minds tell us, and how we relate to the things our minds say, is really very important. It can affect the trajectory and the quality of our life. It can also impact how we live our lives with FSHD.

Let's back up just a little bit more and talk for a moment about the brain. One could call the brain our survival organ. You can also think of it as the orchestra conductor, or a like traffic cop standing in the middle of a busy street directing traffic. You could argue that other organs are more important. If the heart stops beating, or the lungs stop oxygenating blood, we will die. That's certainly true for many of the body's organ systems. All are necessary! Yet, the brain has a role in regulating the function of all of those systems, and nearly all of that regulation is done in the mental background, outside of our conscious awareness. If we had to consciously think about making our heart beat each time, and making our lungs breathe in and out teach time, we would surely die!

Now just think about this for a moment. While you're sitting here reading, your brain is working in the background. It's directing your heart rate, controlling your breathing, receiving vital sensory information from inside your body, and monitoring multiple channels of sensory information from the outside world around you. It does this all in the background, with enough bandwidth left over for you to be able to read and think and carry on a conversation.

As you sit here, some of that information may be analyzed, and deemed to be important, and will get stored as memory. If something that I write in this book seems important to you, you may end up remembering it.

Now pay attention here - this is really important! The brain is amazing, but our brains have limits. The brain can't monitor, pay attention to, and analyze everything inside and outside of our bodies simultaneously. So the brain makes extensive use of shortcuts. It lumps similar things together into groups, and then organizes groups of similar groups into bigger groups. Behaviors that we do all the time, after they are learned - become automatic. They no longer require much attention to perform.

When you watch a toddler – just learning to walk, you can almost see them having to think about where to place each foot, and what to do next in the sequence of getting from here to there. Yet after a few weeks or months, they're beyond walking - they're running – nonstop. Once the brain has learned how to do a behavior that it plans to do regularly, it becomes automated - and we can do it on autopilot without thinking. That frees up space to do other things.

But functioning on autopilot, while efficient, can also get us into trouble. Being able to drive on autopilot - frees us up to do other things like talk to someone on the phone, or worse yet look at our text messages. But if something unexpected happens while we're distracted driving the car, the consequences can be deadly.

The shortcuts that the brain creates aren't perfect, but they're usually useful. Those shortcuts can be specific learned patterns of movement, like walking or running or climbing. Shortcuts can also be "mental rules" for what's usually the best thing to think or do in a given situation. We all operate using multiple mental rules, for how we should conduct ourselves. Rules can simplify things. They can guide our course, keeping us from having to make a specific decision in every single instance. Yet all shortcuts and mental rules are imperfect – they usually save us time, but sometimes they can send us in the wrong direction or fail us completely.

You might've noticed I made a switch a couple paragraphs back. I started to talk about the brain - when earlier I had been talking about the mind. Aren't my mind and my brain the same thing? I guess that the way I'm using the terms in this book - the answer is both yes and no.

The distinction I'm choosing, is to use the term brain to refer to the physical organ in our skull, that interacts with and regulates other bodily systems in the background, and stores memories of our past experiences. I'm choosing to use the term mind to refer to the function of the brain that has self awareness, and that uses internal verbal language to provide a running commentary on our existence – moment to moment.

Analyzing, evaluating, categorizing, and labeling are things the mind constantly does. Imagine two rocks of different sizes, colors, and weights. Imagine one is gray and one black, one bigger, one smaller, and one heavier than the other. If I am looking at the rocks and tell you, "the gray rock is bigger than the black rock," I've provided a way for you to better understand what I am seeing. You might also assume that the bigger rock might even be heavier. Based on past experience, your mind has made a "mental rule" that, in general - bigger things are heavier than smaller ones. Your mind might use this rule as a guide even though it knows that it's possible that a medium size rock could actually be heavier than a much larger bag of cotton balls.

Not only does the mind create rules that don't always work, but language itself can also be confusing. For example, if I tell you that the small rock (black) is "lighter" than the big rock (gray), you might be confused - because I earlier told you that the gray rock is bigger. You may think I was referring to lighter in weight when I actually was referring to lighter in color. Your confusion is due to one of the many peculiarities of our shared language. It uses the same word to describe both relative color and weight. You actually need more contextual information to determine what I mean. Is the gray rock lighter in color than the black one, or is it that the smaller black rock is physically lighter than the heavy gray rock? Sometimes in a rush to quickly evaluate, our minds forget that context matters and make mistakes.

Please bear with me – we're almost there! I can imagine my mind reading this and saying – *Great! Now he's talking about rocks – what does any of this have to do with FSHD?* We are almost there, I promise! While there are many amazing benefits from the mind's abilities to form associations and analyze and make judgements, there are also ways in which those same processes can at times be unhelpful and hurt our ability to cope with FSHD.

Our minds can motivate us to make unhelpful choices, that take us further away from the things that matter most. and we can end up choosing to engage in behaviors that actually increase our inner suffering with this terrible disease. Let's look further at how our mind uses language in ways that both help us and hinder us.

Our minds make mental evaluations about the physical characteristics of things. In the example above, the mind determined one of the rocks was gray and the other black. Our minds also generate evaluations that are less tied to the actual physical characteristics of things in the real world. This kind of evaluation can also be useful within the physical world. For example, imagine I'm a hunter-gatherer, a dad teaching his child how to hunt. If I'm hunting with a sling, I may teach my child that "good" rocks are small and round, and that they fly fast and true toward the prey that I am tracking. There's nothing inherently good about a small round rock, it depends entirely on context. In that context, good rocks are small and round. However, if I am teaching my child how to hunt a larger animal close up and face to face, a "good" rock might be large and heavy with edges that can be shaped and worked so that they pierce and cut flesh and bone.

Evaluations and judgments that are not explicitly tied to physical characteristics can become problematic in our inner world while sometimes useful in the outside world.

These types of mental evaluations that our minds direct towards ourselves, such as good versus evil, weak versus strong, pretty versus ugly, smart versus stupid, normal versus disabled, if believed as objectively true, can feel intensely punishing. They can diminish our motivation to keep moving forward, or worse motivate us to turn away from experiences that could be meaningful and uplifting.

The challenge is that the mind is by nature an evaluation generating machine. However, if we simply accept every mental evaluation about ourselves and hold tightly to them, believing them to be true, we may end up making choices and taking actions that impair our mental health and diminish our quality of life.

Here's the real problem. We respond to our minds, but our minds don't always know what's best for us. Our robust and creatively innovative minds can produce painful unintended consequences both in our physical and inner mental worlds, and not just for us as individuals, but also for society. The fossil fuels we used to power an industrial revolution and improve people's lives now pollute our skies and waters and threaten a looming climate catastrophe. The plastics we developed to make, distribute, and store products, discarded, now fill our oceans and poison sea life.

Our robust and creatively innovative minds can produce painful unintended consequences in our inner mental world as well. The constant flow of our thoughts - imaging, relating, analyzing, and evaluating - when negative and directed at ourselves, can generate painful, unhelpful, and self-destructive meanings and associations. For instance, your mind can retrieve a memory of an unexpected, perhaps slightly uncomfortable encounter with another person. It might then begin generating a flood of evaluative thoughts, *"That was stupid," " Why did I say that?" "I'm such a moron," "Maybe I should just stay away from people altogether."*

A mind can be an evaluation generating machine. It simply does this, nonstop, without our permission. And if we're not careful, if we believe too strongly in the evaluative content generated by our minds, our mental health can be affected negatively. Not everything our minds tell us is useful, helpful, or even true.

So how might the mind impact an individual with FSHD? Let me use a personal example. My shoulders and arm muscles have weakened to the point where I can no longer raise and hold my hands above my shoulders. This means there are certain things I can no longer do - such as safely get a glass from an upper kitchen cabinet, or press a barbell above my head to exercise. My mind can label this inability as being weak. Now, it's important to recognize that with a disease like FSHD that particular mental evaluation is actually quite accurate. The muscles in my shoulders and arms are actually much weaker than those of a man of my same age and body type who does not have FSHD.

The problem comes with all of the other meanings that my upbringing and prior life experiences have linked to the word weak. Maybe as a kid, my own lack of strength and speed made me the last person picked when choosing sides for a sports team. Maybe, when I tripped and fell and started crying, a parent said to me *"Don't be a weakling! Stand up and push through the pain!"* Or maybe later, I saw others looking at my skinny arms and shoulders at the beach, and pointing and laughing.

Experiences like that could've left me associating the word weak with being unwanted, not valuable, and worthy of ridicule. Those learned associations with weakness can then begin to impact how I see myself, and influence how I behave around other people, and perhaps affect the choices that I make about important areas of my life.

Our brains have evolved to make associations in memory based on how things looked, felt, when they happened, where they happened, and based on how other people responded around us. This is an amazing capacity that gives human beings the ability to be imaginative and creative, and invent things that have never before existed. But it's the same capacity that allows a simple word like "WEAK" to be linked with past experiences, and attitudes learned from others, to create personal meanings that are painful and even harmful. And the problem is that memory only works in one direction for the most part. Making these associations (i.e. learning) happens automatically. Yet, forgetting those learned associations once they exist is very difficult short of getting a brain injury or dementia.

My mind might first recognize the actual reality of my FSHD (progressive increasing muscle loss and weakness). Still, then because of past experiences and associations it might go beyond that and also evaluate me as defective, or unworthy. It might tell me, "I am just a quitter" when I experience pain or fatigue following physical activity. If I cling tightly to these ideas as being "true," I might then make choices that are not helpful, and avoid activities that might actually be meaningful. My mind may influence my automatic behavior in unhelpful directions based on a few unhelpful evaluations and learned meanings linked to them. What if these evaluations are just what my mind has learned and simply what my mind does? What if when I'm not paying close attention, my mind is leading me in unhelpful directions?

Evaluative words like "weakness," or "disabled," when associated with painful past experiences and fixed negative beliefs and attitudes, can negatively impact a person's ability to flourish in the world. Whether those beliefs and attitudes are held by other people in the external world and we take them to be true, or whether they arise as evaluation from our own mind, or both. When these words are held to be literally true, and used to guide our choices, they can diminish our ability to thrive in the world and flourish.

I'm going to make a slight detour here, away from the way our individual mind works, to talk for a few moments about how these mental evaluations and language can impact society at large. Humans are social creatures. We don't live our lives in isolation. Most of us grow up in families, tied to a larger community, part of a nation, and ultimately a global world. While our minds generate thoughts internally, some of those beliefs and thoughts get shared, and come to be held as true and taught within communities. However, just as for the individual, not everything that society holds to be true, is useful and helpful. Sometimes those shared thoughts, beliefs, and attitudes can end up producing hurt, prejudice and discrimination, and motivate hurtful behaviors that cause great suffering.

Language about disability in the broader societal context

Words and language matter, not just for individual minds, but to how society at large functions also. Here's another thing. Weaknesses and strengths are not absolute, even in the physical world. When a person cannot lift an automobile or remove lug nuts from a wheel with their fingers to change a flat tire, they don't usually think of themselves as weak, in spite of the fact that their strength is insufficient to accomplish the task. Rather, that degree of weakness is assumed, so much so that automobiles are sold pre-equipped with special assistive devices (jacks and lug wrenches) for changing a flat tire.

Yet, until recently, homes and businesses have been designed that failed to consider the needs of individuals who lack sufficient strength to climb stairs, or open heavy doors. It has required Congressional action and vigorous enforcement of laws to begin to ensure that buildings (and bathrooms) are accessible to everyone. This process is still ongoing and often resisted.

Mostly people are oblivious to the impact that physical ability differences can make on quality of life and meaningful living. As I mentioned, I've had symptoms of FSHD for close to 50 years now, but it's only very recently that I've begun to think of myself as disabled. There was a time in my career just 15 years ago when I was traveling extensively by air. The physical requirements of getting through airports, boarding, and flying to and from my destination were automatic. I never had to think much about travel time or where I was seated. It was all just about getting there and back.

More recently, I began to need certain accommodations. I would have to request a wheelchair to ensure that I could get to the next gate in time to catch the flight. I now use an electric wheelchair that I take down to the plane's door, where it gets stowed in the baggage compartment. Still up to this point, though, I can walk onto the plane, down the aisle, and find my seat. Even with my own challenges, it never really occurred to me how the experience of flying is for others with different abilities. That finally changed for me when I came across a Facebook post written by a young woman I had met and talked with at several FSHD conferences.

She is now wholly confined to her electric wheelchair and currently cannot stand up from a chair or walk even short distances. Her Facebook post was a moving and powerful statement of gratitude for the kindness of strangers. When she flies, she must refrain from eating or drinking for 6 to 8 hours before boarding. This is because once she has been seated in the aircraft, using the special narrow wheelchair that can fit down the airplane aisle, she is stuck there until airport personnel at the destination enter the plane with a similar wheelchair to take her out of the aircraft. As it happened on that particular flight, her pre-flight efforts failed, and she needed to use the restroom. She wondered what she was going to do until a kind, strong man offered to help. He lifted her up from her seat, carried her piggyback down the aisle. He waited with her on his back, and then helped her into the tiny bathroom when it became available. A female flight attendant helped her stand and pull up her pants so the man could carry her back to her seat.

What struck me so overwhelmingly about her post was that she accepted her physical condition and that her first and most prevailing feeling was one of gratitude for the kindness of strangers. My response to the post was to feel rage at the culture that pretends it is OK to design transportation systems for long journeys that physically prevent certain individuals from using the bathroom. However, her willingness to hold her natural emotions of anger, embarrassment, and humiliation lightly has allowed her to travel worldwide, have memorable experiences, and feel gratitude toward kind strangers.

If she were to hold tightly to those natural feelings of embarrassment and anger and avoid air travel entirely, she would lose out on incredible opportunities for new experiences, as well as positive emotions such as gratitude. Her natural anger at society’s lack of awareness of the needs of the disabled has provided benefits also. She has become an influential advocate for accessibility within her local community. Her strong voice in her community has led to structural changes that have improved access within that city and benefited thousands of other people with mobility challenges.

Language and evaluation are a core part of how our minds work. Words and evaluations arise from within our mind, and when those words and evaluations are shared verbally with other people, their impact moves beyond the individual and shapes the thinking, attitudes, and behavior of the larger society. We also are shaped and influenced and learn from the language of the people all around us, families, friends, and teachers.

Acceptance and Commitment Therapy targets language. It teaches us to notice the verbal content all around us, and provides more workable ways to engage with the verbal content, the memories of past relationships and experiences, and the emotions that flow from our minds. The relationship that we have with the thoughts, emotions, and memories that our mind generates matters. How we choose to engage with our inner world makes an important impact on our overall mental health, satisfaction with life, and happiness.

The Problem with Control and Avoidance

Avoidance and control have been two strategies that human beings have used across millennia to thrive in the physical world. Human survival is in large part due to avoiding or controlling the many threats to physical existence. Small children are taught by their parents the dangerous things to avoid in life. Parents create simple rules for their children to follow to stay safe. *"Don't touch the stove. It's hot," "Don't run into the street, you'll get hit by a car," "Never get into a car with a stranger."* We also learn to control our environment to relieve physical discomfort. If the house is cold, either turn up the heat, put on a sweater, or curl up with a blanket. If you're hungry, go find something to eat. If your head hurts, take medications.

We learn these strategies as children by listening to our parents' instruction and observing the efforts of other people. And these strategies work effectively, as a rule, to keep us safe, comfortable, and satisfied in our physical environment. Children learn to wear warm clothes when it's cold outside, to put on protective gear to prevent injury when skateboarding. Sometimes that is learned directly through parental instruction, and other times through painful previous injury.

Many children also get instructed to try to exert similar control over their inner experiences – their thoughts and emotions. Frustrated parents may say things like *"Stop crying or I'll give you something to cry about!"* This teaches the child that certain emotions are inappropriate and should be controlled or avoided.

It's not a surprise then, that our natural inclination, our immediate response to experiencing unpleasantness in our internal world, is to utilize the same avoidance and control strategies that work so well in the external world. When unpleasant memories, painful thoughts, or difficult emotions arise within us, our first inclination is often to try to avoid having them, or to exert control over them in some fashion. It makes sense, since these strategies work every day for us in our physical environment, why wouldn't they work inside of our mind and body as well? It wouldn't surprise me right now if your mind was telling you that I'm crazy, and you shouldn't believe me. I can tell you that there's actually a great deal of research evidence that suggests that what I'm saying is correct. Attempts at control over our internal thoughts and emotions can even be paradoxical. You still might not believe me so let me just give you a quick example of what I'm talking about.

Remember the childhood rhyme about the purple cow? *"I never saw a Purple Cow, I never hope to see one, But I can tell you, anyhow, I'd rather see than be one!"* (Gelett Burgess) I'd be willing to wager that most of you reading this haven't thought about a purple cow for many years, maybe since the last time you heard that poem. But if right now, I were to instruct you – no matter what - don't under any circumstances think about a purple cow – not even one time - for the next 10 minutes. Go ahead – try it. Try not to think about something specific. See if you can do it.

What you will most likely discover, is that in order to make sure you're not thinking about the cow, your mind has to bring the purple cow to mind to compare. Research suggests that when people try to not think about something, there's actually a rebound effect and they end up thinking about it more than they would have before they tried not to.

Now, not thinking about purple cows is a silly example – trying not to think about them doesn't have any real costs associated with it. However some efforts we make to avoid unpleasant thoughts or emotions about our FSHD can actually add to our physical suffering, and reduce the quality of our lives. If I'm out in public, and I experience unpleasant thoughts that "*I'm ugly and disfigured*," and "*people don't want to have to look at me*," I may decide to stop going out in public. And there's a cost to that avoidant choice. It keeps me stuck at home, prevents me from doing things that I love or need to do. It keeps me from being with people who care about me. Or, maybe I decide to respond to my painful emotions of sadness and loss because of my physical decline, by drinking or using substances, and eventually over time I now have two problems – FSHD and substance abuse.

The world of advertising is filled with subtle (and not-so-subtle) instructions to teach people to try to control their inner emotional experience (by buying certain products). If you're feeling sad or down "talk to your doctor" to get a prescription for the latest anti-depressant or anti-anxiety medication. If you're shy and lonely – commercials suggest - buy a beer and you'll be instantly surrounded with happy people that love you. It must be true, you watch it happen in the commercial! If you feel alienated and rejected and want people to like you - simply purchase the latest fashions and you'll instantly be a part of the group. The reality is, however, that trying to control our emotional experience through purchasing is a short term, and ultimately unworkable (as well as costly) exercise.

Spending money to purchase products is not the only strategy we use to try to control our unpleasant thoughts and emotions. Avoidance is the strategy I used frequently when I was growing up. For the bulk of my late elementary school and on through high school I was painfully shy. I wasn't very popular, and if the teacher called on me in class - my face would turn bright red, and classmates would point at me and laugh. I would do almost anything to keep from having that experience or try to shorten it. I would never raise my hand to participate in class, and if called on by the teacher to answer a question, I would immediately say "I don't know," in hope that it would take me out of the spotlight more quickly.

Those efforts at avoidance weren't very effective, and had the added cost of worsening my class participation grades. The more I tried to isolate myself and avoid ridicule, the smaller my social circle got. That meant I had fewer supporters, and more people outside of the group that viewed me as an outcast. My experience with shyness didn't begin to change until I began to take a few risks. I had to be willing to risk embarrassment, and talk to other people. Only then did I discover, that not everyone would laugh when I talked. Ultimately, I began to learn that some people could appreciate what I had to say, and in some instances even look to me for leadership.

Living on autopilot can lead to inflexibility

I mentioned earlier our brains look for ways to make certain behaviors automatic. When our brain identifies a behavior that we tend do a lot, the individual parts of those behaviors, tend to get clumped together, such that the whole chain of little behaviors can be done in sequence without thinking. As an example let's consider driving. Each of us probably retains some memories related to learning how to drive. For me, I remember moments of feeling almost overwhelmed by the sequence of required tasks. There's a whole series of behaviors related to getting the car started. Sitting down, adjusting the seat to the right distance from the pedals, checking the mirrors, making sure the car is in park, then inserting the key to start the engine. Now, recent innovations and technology have eliminated the need for some of those things.

Once the car is started, we start a whole new chain of behaviors. We place our foot on the brake, shift into drive, glance around in all directions, move our foot to the gas, and begin to move forward. As a brand new driver, the constant monitoring, and worrying about making mistakes is anxiety provoking, and feels all consuming. But within a few weeks or months, the process has become automatic. Suddenly, all of that stuff is now happening in the background, while you are actively engaged in conversation with friends, or listening to (and maybe singing) your favorite song on the radio.

However, at times automatic behavior can become a problem. Some of us may have had the experience of being so engaged with something else while driving, that we missed our exit on the freeway, or suddenly realize that we have no recollection at all of the cars that were around us or the lane changes that we made, while we were busy thinking about something else.

Avoidance can cause us to become inflexible related to FSHD as well. Automatic ways of responding work well in situations that are safe, predictable, and unchanging. But FSHD is not like that. Subtle changes can be happening within our body that can suddenly make once safe activities - unsafe. Problems can arise when something suddenly changes and our usual (automatic) ways of doing things no longer work. I tend to think of inflexibility as persisting or continuing in a behavior in spite of direct evidence that it's not working for me or helping me.

Instead, often I find (usually by reflecting after the fact) that I'm continuing this behavior because it helps me avoid some other internal experience, such as emotional sadness or loss. Sometimes painful thoughts about what stopping the behavior would mean about me, makes me stubbornly refuse to change. Most often I'm not even truly aware of the fact that I'm continuing to do things that aren't helpful. The habit that I have formed of doing something in a particular way, keeps the activity outside of my conscious awareness. With FSHD, inflexibility or continuing to do things that can potentially harm me can be costly.

The costs and consequences of being inflexible and using avoidance and control strategies within our inner world of thoughts, sensations, and emotions are variable and highly individual. There may be certain circumstances where avoidance and control is not a problem. This book is not trying to get anyone to change those things that are truly working for you. The focus of ACT is to help you to begin to notice when using inflexible avoidant strategies **IS NOT** working for you- and to provide you with workable alternatives. The remainder of this chapter will be devoted to exploring more fully the mental and behavioral processes that are a core part of Acceptance and Commitment Therapy.

Opening up

The first leg of the three-legged stool of ACT is the practice of opening up. This practice has two distinct core processes – acceptance and defusion. These processes can provide an alternative to the sometimes unhelpful strategies of avoiding or trying to control our inner experiences such as disturbing thoughts and emotions.

Human suffering and additional struggles arise when we try to apply these avoidance and control strategies to our inner world of thoughts, emotions, sensations, and memories. ACT suggests that acceptance and defusion provide an antidote to the unworkable strategies of avoidance and control when engaging with the unpleasant content of our mind and our inner world.

Imagine that you are sitting in your office one day, surrounded by bookcases and filing cabinets. It's a fancy office, and there is a large chandelier hanging from the ceiling above your head. One day, without warning, there's a powerful earthquake. You sit there at your desk for a second or two, trying to comprehend what's happening. Then you realize you're supposed to do something, and you scramble quickly to shelter under the desk. As the shaking continues - books fall, drawers in the filing cabinet pop open, the chandelier crashes down onto the desk, shattering glass - shards fly everywhere. You're terrified, afraid you're going to die, as a part of the ceiling comes crashing down.

Afterward, you are rescued, go home, and life goes on. Over the weeks and months following the earthquake, construction crews fully repair the damage, and your office looks just like it did before. If someone were to come into your office, they would have no way of knowing what had happened. Someone else could move into the office and have no fear, no anxiety. Buildings don't have memories. Unless someone posted a newspaper clipping about the earthquake on the office wall, the new occupants might never know. In the physical world, there's no easy way to go back in time. Past events are past. Only the present remains.

However, in the inner world of our mind, it’s a different story. There, many past events live on in our memory forever. Unless serious brain injury or dementia takes it away, that fearful memory of your earthquake experience lives. The memory of that fear can re-emerge suddenly, “without permission,” when something reminds you of that past event. I mentioned earlier our brain’s ability to make associations automatically. When a significant event occurs, that event is automatically associated with the experiences that were present in that moment. The sensations and emotions are linked together in memory, and often those memories will come into awareness in the presence of reminders of those things.

So when you walk back into your office and it looks like it did at the time of the earthquake, you may feel the same intense emotions experienced during and after the earthquake. Whatever evaluations or judgments you made about yourself and your actions remain as well. Those arise too. If you labeled yourself a coward, if you told yourself you just weren't any good in a crisis, and if you believe those thoughts – it may shape your behavior in the present and future. If the recurring emotions, sensations, and thoughts continue to feel intensely painful, it may affect your ability to work. You might begin to avoid reminders of the experience. Offices might make you uncomfortable. You might quit your job and give up a career you love – all to try to avoid and control your painful thoughts and emotions about events long past, that no longer exist in the present moment.

As a clinical psychologist who specialized in posttraumatic stress, I have interviewed hundreds of people haunted and impacted by traumatic experiences in their past. However, individuals with FSHD can be similarly affected by sad memories of activities in which they can no longer participate. They can be haunted by discouraging or hopeless thoughts about their physical decline, and filled with worry and anxiety about their future. Having painful thoughts, memories, and emotions is not the real problem. Having those experiences is normal. What can become problematic are the ways we choose to engage those experiences. Trying to avoid and control experiences that are outside of our conscious control, can be both costly and unworkable. Trying to avoid and control painful mental experience, can end up actually increasing our suffering.

Now that we set have set the stage by looking at some of the costs and consequences of avoidance and control, let's begin looking at the alternative – acceptance.

Acceptance

The first of the two processes that make up the practice of opening up is called acceptance. The following is a definition of acceptance from an ACT perspective:

> *"Acceptance, as we mean it, is the voluntary adoption of an intentionally open, receptive, flexible, and nonjudgmental posture with respect to moment-to-moment experience. Acceptance is supported by a "willingness" to make contact with distressing private experiences or situations, events, or interactions that will likely trigger them."*[5]

Often when people hear the word acceptance used - what comes to mind is some sort of passive resignation to the worst experiences life brings. People often hear acceptance as giving in or giving up in the face of difficult external problems. That is not it all what we are talking about here. Rather, we use acceptance to describe a new stance we choose to take with regard to our internal mental and emotional experience. Acceptance is about how we choose to respond when difficult, painful thoughts, emotions, and memories show up and begin to take up residence in our mental experience. Acceptance is an alternative way of engaging with this mental experience that stands in contrast to the unworkable strategies of avoidance and control that we have used in the past.

When I was in grade school, one of my favorite activities at recess was to swing on the swing set. I enjoyed that feeling of rushing forward to new heights - seeing just the sky at the peak of the swing and then falling back again until, at the other end of the arc, I am facing the ground. Over and over, back-and-forth, with feelings of excitement and joy. One can imagine as a child wanting to stop that experience of the swing at the perfect spot, perhaps right at the top, to never have to leave that place. But we know that's not possible.

Empty Swingset

Just as the swing moves forward to the top of the arc, inevitably gravity takes over and pulls us back again, over and over, back-and-forth, until eventually, the swing stops. Playing on a swing set is an ever-changing process. Effort that we exert to start the swing pushes us forward as far as possible, and then gravity pulls us backward, and the process continues over and over. If we continue to exert any energy at the right time, we will swing higher. Once we stop exerting energy, eventually (but not immediately) the swing will stop.

There are ways that our inner world (emotions, thoughts, memories, sensations) is like a swing set. Think for a minute about emotion. Emotions show up in our awareness and increase to a certain peak intensity. Then, at some point, they diminish and fade away out of awareness, only to return at some later unexpected date. Humans have little conscious control over the rise and fall of our emotions. Now, emotions don't appear randomly. Experiences or interactions trigger emotions in the physical world. Our thoughts, bodily sensations, and memories also trigger them. Our inner world experiences are linked in complex ways.

Thoughts are similar. Thoughts pop into our minds, and then fade, replaced by other thoughts. The difference is we have some intentional control over our thoughts. I can decide I'm going to think about a particular thing. However, even then, some other random thought will show up and replace it. Random thoughts get triggered by memories, associations, or things occurring in the physical environment that enter our inner world through our senses.

We all know that some inner experiences are unpleasant. No one wants to be overwhelmed by sadness or feel the discomfort of strong anxiety. Acceptance does not mean that you WANT to have painful experiences. Acceptance acknowledges that both painful and pleasant inner experiences occur and that trying to avoid them or engaging in actions to control those internal experiences is often ineffective and counterproductive. It may create unintended and often worse consequences, such as prolonging or intensifying the experience.

When speaking about acceptance in a group of people who have FSHD, some who initially hear the word acceptance understand me to be suggesting that people simply “give in to” or “give up” to FSHD. They tell me of their powerful need to continue to fight the disease. To clarify, acceptance does not mean giving up–or failing to do everything possible to treat, manage, and cope with the physical impact of FSHD. Instead, acceptance has to do with how I choose to engage with my internal mental experience of the disease. It refers to how I engage with my thoughts, emotions, sensations, and memories.

If the word acceptance feels problematic, another word for acceptance used in the ACT community is willingness. Being willing doesn’t mean I want to have these uncomfortable feelings and thoughts about my disease and about myself. Rather, willingness is an acknowledgment that efforts to avoid or control those inner thoughts and feelings rarely work very well. Willingness is being intentionally open to the natural shifts of your emotions, thoughts, memories, and bodily sensations when they show up in your awareness. Willingness means engaging with those experiences, lightly, without struggling, knowing that even if unpleasant, they will at some point fade again out of your awareness if you don’t hold on to them.

During 2020, I was interviewed on the FSHD society internet radio podcast. In that conversation with Tim Hollenback, the radio host, I spoke a bit about ACT and briefly described some of the key processes. Based on some comments received on the chat stream, the word acceptance was impeding people's ability to grasp that ACT process. As I spoke about acceptance, some people seemed to hear it as "giving up" their struggle with the disease. They strongly resisted the idea of giving up. During that brief radio conversation, there was insufficient time to fully explain how an ACT understanding of acceptance differed from giving up. I hope I am making that distinction clearer now as I write this book.

In ACT, we do not direct acceptance at a physical disease process that manifests in the physical world. Instead, acceptance is about the relationship I have with my inner emotional and cognitive (thinking) experience. Acceptance is NOT about abandoning my external physical battle with the physical disease FSHD. It means acknowledging and and being willing to experience whatever emotions, sensations, and thoughts show up inside me about the disease. I notice the thoughts that emerge in my mind. Whatever emotions show up in my body, I notice the sensations connected with those emotions. Whatever internal experience shows up, I let them be as they are, without evaluation. I let go of my desire to struggle with or keep from having those feelings, sensations, and thoughts. Acceptance is an alternative to experiential avoidance. Research suggests that when I actively try to avoid or stop having certain thoughts or emotions, those efforts are usually unsuccessful and often increase the intensity and duration of those uncomfortable inner experiences.

Let me give you an example from my own FSHD experience. In the late 1990s, the FSHD disease process moved into my legs and feet. I noticed I was walking much more slowly, and I started stumbling and falling more often. The VA hospital where I worked closely cooperated with Stanford University. So I sought an appointment with a Stanford neurologist. After evaluating me, one of his recommendations was that I get AFOs (ankle-foot orthotics) to prevent the foot drop that was causing me to stumble and fall. He gave me a referral to a prosthetics company that could provide the AFO.

I didn't act on that prescription. The idea of the AFO bothered me. Thinking about wearing one made me feel afraid that the disease was winning. I had thoughts that wearing an AFO would mean I was becoming disabled, and those thoughts felt really uncomfortable. So I simply avoided doing anything about it. I convinced myself that my shoes were the problem and that I simply needed to find the right shoes so that I wouldn't stumble. I spent a lot of money on shoes over the next few years. The soles couldn't be too slippery or too sticky. The soles couldn't be too thick or the shoes too heavy.

My attempt to keep from having emotions of sadness and fear and to avoid coming into contact with thoughts that I was becoming disabled wasn’t really working. Shoes weren’t the problem—my disease was the problem, and my thoughts and feelings about FSHD were keeping me stuck. I continued falling - for years. Once I fell while standing and speaking in front of a group of 40 patients. I turned to write something on the whiteboard, and suddenly I was down on the ground. Every one of my close friends during that time had the experience of helping me up off the ground after I fell mid-sentence while walking and talking with them. My attempts to keep from having sadness, fear, and uncomfortable thoughts about disability were not working, and it was harming me physically. My elbows and knees were battered and bruised. I stopped walking with people, and my physical activity decreased. I got physically weaker. My efforts to avoid internal emotions and thoughts were hurting me in the physical world. Those efforts were completely counterproductive - they brought unintended consequences. Trying to avoid internal mental discomfort —was actually hastening my physical decline and worsening my physical weakness.

It took me seven years to become fully convinced that my life would be better (and my bruised knees less painful) if I just got the AFOs. But to make that move, I also needed to be willing to have whatever thoughts and emotions might come up. Finally, at long last, I went to the prosthetics clinic and get first one and then ultimately two AFOs. My AFOs became an asset in managing my disease, not a sign of my giving into it. Years later, I had a similar battle in my mind over the need to use a scooter and wheelchair. I now realize that emotions like sadness or fear of disability will continue to show up from time to time. However, I also realize that I can make intentional choices about how I respond to them.

My mind hangs on to the things that I learned and observed growing up in this society. I still carry inside me the implicit bias about disability that views people wearing leg braces or being in a wheelchair as weak, less capable, and less worthy. However, I also know that by trying to avoid painful emotions and thoughts that arise around using a wheelchair, I stayed stuck. My efforts to avoid having these painful thoughts and emotions created consequences that made my life worse. When invited to go shopping or with friends on an outing, I would choose to stay home because the walking was too difficult. Trying to not have painful thoughts and emotions caused me to miss out on chances to be with people I care about and to have fun new experiences. When I opened myself to having those feelings and thoughts, and got a wheelchair to use when out and about, I experienced new opportunities for joy and social connection. I wondered why did I wait so long? Willingness (Acceptance) was and continues to be a crucial element in my fight to have and maintain a life that is worth living.

Hank Robb is a psychologist that has written about willingness.[6] He uses a simple mental exercise similar to this one to explain willingness.

* * *

EXERCISE 2.1 Holding a Cactus

Take a moment and extend your hand in front of you with your palm up as you read. Imagine that you are holding a small white feather in your hand, much like in the picture below. In your mind, imagine holding the feather gently, in a way so that it is undamaged. Pay attention to the experience of holding the feather. Notice its softness and how light it is. Notice that you can hold it with tenderness and kindness. Continue simply imagining holding the feather for a moment, paying attention to that experience.

Feather

Now imagine that instead of a feather, you are holding a cactus. See in your mind the sharp spines extending in all directions. The cactus doesn't feel soft but prickly in your hand. And yet, notice that you can hold it just as gently, and with the same kindness as you did the feather. In fact, holding the cactus gently will actually make the experience less painful.


Cactus

* * *

If you were to grasp the feather tightly in your fist to control it, you might damage the feather or destroy it. If you grasped the cactus tightly in your fist, the experience would be quite painful. Our inner world is similar. Efforts that we take to avoid or control our inner experiences are often unproductive and may generate even more pain.

Defusion

The second necessary process engaged in the practice of opening up is called "defusion." In order to understand defusion, it may be useful to better understand what we mean by its opposite - mental fusion. The ability to generate creative ideas is one characteristic of minds that has helped human beings survive in the world. For example, I can use my mind to envision and then design a tool that, if built, would be extremely useful in my physical world. However, until I use my hands and feet to get the raw materials and then to shape and actually build the tool - it's not real - I can't use it. It's only a mental construction.

Let's try a real example using a metaphor called "Finding a place to sit," described by Steven Hayes and colleagues.[7] Let's say you're tired after a long day and you need a place to sit, and so you begin verbally describing a chair. You give an incredibly detailed description of a chair, its color, the fabric, the design of the frame. Your description is so detailed you can picture it in your mind - your verbal image of the chair seems so real that you could reach out and touch it. But remember, you're tired. Can you actually sit in it? NO! Verbal description and analyses are fine - there's nothing inherently wrong with them. But when we're tired and need a place to sit, they're not helpful. Verbal knowledge and actual experience are two different things. You can't automatically substitute one for the other.

Check your own experience. My mind provides a near constant narrative - telling me that my world is this way, and that my problem is this thing. It describes my situation and evaluates how I'm doing constantly. In addition, my mind will frequently provide "rules" about what I “should” do. My mind derives these rules from my experiences or through observing or being instructed by others. However, all the descriptions and evaluations my mind generates are only helpful and workable when they mesh with my actual lived experience in the present moment. Sometimes, the verbal descriptions, evaluations, and rules that my mind provides may simply keep me rigidly going in a direction that's not working. I may find that my mental fusion makes me inflexible, and keeps me from responding to the present moment in ways that are consistent with my most important values.

If I'm tired, descriptions and evaluations of imaginary chairs are not helpful. Rather, the lived experience of dropping my body into an actual chair and feeling my muscles actually let go of the struggle required to stand is what I truly need to find relief. When I actually sit in a real chair, I can relax and allow the chair to support my weight instead of my muscles, and I can experience genuine relief. My mind’s most detailed mental construction of a chair can't replace my actual lived experience.

So, defusion is a skill that I can practice. It is a skill that can help me to "de-literalize" the verbal content my mind is providing. I can use it to step back and see the bigger picture. Defusion can provide me the space to not simply follow my mental rules on autopilot, but to choose intentionally to respond in the present moment in a values-consistent way. Defusion is a skill that can enhance my psychological flexibility.

If fusion is entanglement in thoughts, defusion is the process through which we become less entangled and free ourselves. Our internal mental content does not always conform to the actual reality in the external world. The evaluations and judgments that our minds make about the meaning of events and even about ourselves may not be literally ”true.” However, believing those evaluations and acting upon those beliefs can sometimes wreak havoc in our lives. John Blackledge writes, *“Within ACT, defusion techniques involve a variety of actions designed to expose thoughts simply as thoughts rather than binding realities.”*[8] Cognitive defusion techniques help us perceive our thoughts as thoughts and hold them lightly. This gives us the space to choose our next actions intentionally, to decide which are most consistent with our values, unaffected by our thoughts' content.

One goal of defusion is to help us become observers of our own thoughts. We must practice defusion to become skilled. The ability to pause and notice the stream of thoughts and images flowing in our minds is a skill that requires effort to develop. One way to develop this skill is by engaging in mental exercises similar to the one I am inviting you to try below.

EXERCISE 2.2 Leaves on a Stream

Close your eyes for a moment. Picture yourself sitting on a boulder beside a tree-lined stream. Watch as the water moves slowly over rocks, around a submerged tree trunk, flowing gently down into a distant valley. Listen to hear forest sounds, birds chirping, the wind rustling through the trees, the gentle sound of water splashing up on the rocks.

Forest Stream

Once in a while, imagine that a large leaf drops from a tree overhanging the water. Notice it floating gently down on top of the water, beginning its slow journey down the meandering stream. Just imagine that you're sitting in the sunshine, watching the leaves float by one-by-one. Now turn your attention toward your thoughts. Each time a thought arises in your mind, imagine it written on one leaf floating in the stream. The thought might be a word or phrase —imagine it written on the leaf floating gently downstream. The thought might be an image —picture it on the leaf slowly drifting away. Your goal is simply to observe - notice what shows up. If the image disappears and you find your mind has gone some place else, notice what it is thinking and put that thought on a leaf, then simply returning to the stream and continuing to observe. Watch each thought come into your mind, placing it on a leaf atop the stream, and watch it float gently away. Continue this for several minutes, then take some time to reflect on the experience.

* * *

I would encourage you to use the above exercise not as a onetime experience, but as a discipline that you do regularly as a way of strengthening your ability to notice your thoughts simply as thoughts. Engaging in this mindful noticing builds awareness that thoughts come and go continually. They help us realize thoughts are a product of our mind that we can step back from and observe. The ability to step back and notice thoughts as thoughts can open up mental space for us to realize there is a self that is separate from our thoughts. Having opened up space, we can then take advantage of our ability to make intentional choices about our actions in the present moment. We don't need to live life on autopilot simply following the mental rules our minds provide. Rather, we can step back and take a moment to notice whether our thoughts are leading us to engage in valued activities, or directing us further away from things that matter. Defusion can help us turn off our autopilot, and become more intentional in the choices that we make.

Another goal of defusion is to disrupt the associations between language and experience to de-literalize the words. Let's explore this further with an example. When I was a child, there was nothing better than an ice-cold glass of milk on a hot day. I have memories of coming into the kitchen, all hot and sweaty, after playing outside. Opening the cupboard door, I remove a glass and set it on the counter. I can see myself opening the refrigerator and taking out the carton of milk. I open the carton and pour a tall, full glass of ice cold milk. In my mind, I remember the sensations of lifting the cold glass to my lips and drinking. I can almost feel the sensations of the cold milk in my mouth, and the satisfaction associated with gulping it down. The word milk has become associated in my mind with pleasant childhood memories from hot summer days in the past. But in reality, the word milk is nothing but a word. The following exercise helps you come into contact with that reality and to see how language gets entangled with memories and learning.

To begin the exercise - you might make sure that no one else is around—so you won't feel embarrassment. If you'd like to practice two core ACT processes, i.e. defusion and willingness (to have the emotion of embarrassment) simultaneously, you could consider doing this exercise with other people around. No matter how and when you choose to do the exercise - hopefully, you're not lactose intolerant. This exercise is based on insights first described over 100 years ago by early psychologist Edward Titchener.[9](p425) Steven Hayes introduced an exercise similar to the following in his first book on ACT.[10]

EXERCISE 2.3 Milk

To begin - say the word "MILK" out loud. Now picture in your mind the scene I just described. You're holding in your hand an icy cold glass filled to the top with milk. The milk is so cold you can feel the condensation forming outside the glass.

Glass of Milk

Hold the glass to your nose and smell. Maybe you feel the coolness from the milk entering your nostrils. What memories do you associate with the scent of milk? Notice as you focus on the image how real it becomes. You can almost feel the coldness from the milk in your nostrils and imagine the taste as you slowly drink and swallow. Hold that image in your mind while you say slowly out loud, repeatedly, the single word MILK. Now repeat it, and again, not stopping. Milk… Milk… Milk… continue saying the word faster, and faster, and then faster still. Notice what happens as that same word comes rapid-fire out of your mouth over and repeatedly. Perhaps you notice that at some point, the image of the glass drops away, and the word that you're saying sounds nonsensical, like gibberish. Suddenly, the word milk has lost its meaning. The effect isn't permanent—stop saying the word, and bring the image of the glass back to your mind—there it is again. At their core, words are just sounds—we provide and attach meaning.

* * *

Humans hold words to be literally true all the time. Our minds engage us in continuous narrative—commenting on and evaluating our moment to moment experience internally. We become so used to our inner voice that we stop noticing that sometimes it may motivate us to make unhelpful choices. Getting caught up in our thoughts may lead us to react by taking actions that make our lives less workable.

With a disease like FSHD, there are many areas where our thoughts can affect how we live in the world. Let's look at a few examples. FSHD affects the muscles in the face, arms, torso, and legs—in ways that change our facial expression, posture, and movement. The disease reduces our physical strength, making us physically weaker than we used to be, and often weaker than other people. The disease causes varying amounts of physical pain and fatigue. Our mind notices all those things, and comments on and evaluates those physical experiences that are part of FSHD. The things that your mind tells you about your disease may differ greatly from what my mind tells me about my disease.

To illustrate, I'd like to walk you through what happens in my mind. Just as an example, imagine the thought, "I can't do that!" With my FSHD, I have developed a constant awareness of my increasing physical difficulty in engaging in everyday activities. Some actions performed easily months or years ago I can now do only with incredible difficulty.

I mentally evaluate every activity I consider engaging in for whether I can or can't take part. If I enter a building for an appointment and look at the directory and find that the office I'm looking for is on the second floor, and I see a tall winding staircase right in front of me, the thought "I can't do that!" may emerge. Along with that thought comes an emotion such as anxiety or discouragement. I may look rapidly around to see if I see a sign for an elevator. If I don't see one - then it's decision time. In my current physical condition, that thought "I can't do that" is not literally true. I CAN climb stairs, albeit slowly, with great effort, sometimes requiring both hands firmly on the handrail pulling me forward. The effort required may mean that I will have to sit down when I'm done. I may also have to be willing to feel embarrassment at being seen by others struggling to do something they find simple. I may have to contend with thoughts of worry that going up the stairs will exhaust me so much that I might increase my risk of falling when coming back down the stairs.

If in looking around, I find the elevator—then problem solved, fear averted, all is well. Except that now, maybe I've strengthened my belief in the literalness of that thought, and avoided engaging in physical exercise. Now, when I think about going somewhere, I try to imagine in advance all the obstacles that I might face. Maybe instead of welcoming the anxiety and holding lightly the literality of the thought, "I can't do that," I might simply decide to avoid the problem, skip the appointment, and stay home. I then become more and more housebound. I stop doing things with people and become isolated. When I do this, I am giving away opportunities for meaningfulness and joy in the present, by going to unfamiliar places and having unique experiences.

The challenge of FSHD is that in some moments, with some activities, my mind's evaluation "I can't do that," may be literally true, and because of disease progression I might lack ability to safely undertake an action (at least without some adaptation or assistive device). Yet, at other times, that same thought may arise to avoid being reminded of the disease's impact. You will need to assess for yourself whether fusion with your thoughts is keeping you from acting or if you physically cannot safely engage in an action. This is the challenge for each of us with FSHD, to become skilled observers of our inner thoughts, and build awareness of our own tendencies to avoid certain situations that bring up uncomfortable thoughts and emotions, without taking risks that physically endanger our health. Finding that balance from moment to moment can help us live with vitality and meaning.

Your mental experience with FSHD might be like mine, or it might be quite different. Take a moment to reflect. What are the thoughts that keep you from doing things that matter to you–things that you would like to do? A person's experience of FSHD is never static, the disease keeps progressing. There's no universal right or wrong answer I can share. The question becomes whether my thoughts are leading me to take actions that make my life more meaningful and significant, or actions that make my life less workable over the long term.

Acceptance and defusion can be powerful tools for experiencing a meaningful life. Practicing these processes in everyday life is a way forward.

I hope that you will experiment with these processes of acceptance and defusion and see what happens as you practice and build your skill with these processes. It will take energy and effort to step back from learned automatic habits of how you usually engage with your thoughts and emotions. Take the time to notice where fusion with your thoughts keeps you stuck in life, unable to move toward the things that matter. Pay attention to moments when you are non-accepting of the emotions that show up within you, and the efforts that you then engage in to push away and not have those emotions. Look at the costs that may come with avoiding these feelings both at the moment and over the long term.

There are additional acceptance and defusion exercises to be found in each of the modules that follow this section. There are many books written about ACT that will also provide additional exercises to explore. I encourage you to use these types of exercises to step back from patterns of avoidance, and from difficult thoughts keeping you stuck. I hope that these and other exercises within this book will empower you to identify your most important values and to take actions to express those values in ways that make your experience of living with FSHD more meaningful.

Awareness

The second leg of the three-legged stool of ACT is the practice of awareness. This practice also has two distinct core processes. These processes help me become more aware of what is around me in the current moment and notice a larger sense of myself.

Present Moment

ACT is one of several modern therapeutic models that places strong clinical attention on the present moment. From an ACT perspective, the present is all that exists. The past is gone, and the future is yet to be written. The present is the only place where we can live life with vitality and fullness.

There's nothing wrong with revisiting treasured moments from the past through memory, nor with making realistic plans and setting goals to be attained in the future. However, key features of many psychiatric disorders include getting stuck in ruminations about past events and decisions. They can hopelessly lead you to spin your wheels with worry about things that might happen in the future. Every moment spent mentally reviewing the past or worrying about the future is a moment lost to living and taking meaningful action in the present moment.

It's very easy to get lost in thoughts about the past or future. Returning one's focus to present moment awareness is a skill that you can develop and practice. One tool used in ACT to help develop the ability to focus attention and to perceive the possibilities available in the present moment is called mindfulness. Mindfulness is a discipline that you can cultivate and must practice. There are many ways you can practice mindfulness. But it takes effort. I've found that the effort is worth the payoff.

Perhaps this metaphor will help you to see what I am trying to convey. For much of my life, bicycling was my primary means of exercise. At one point, I was riding my bicycle over 100 miles per week and took part in several rides of over 100 miles in a single day. I didn't just start out one day and decide I would ride my bike 100 miles. Getting to that point required a long period of training. It required riding longer distances day after day and week after week. It required riding in varied terrain, up and down hills, riding at altitude, and riding against the wind. Mindfulness is similar because the skill is best developed by starting slowly and patiently, building strength and endurance through daily and varied practice.

Jon Kabat-Zinn defines mindfulness this way: *"Paying attention in a particular way: on purpose, in the present moment, nonjudgmentally."*[11] Notice the elements of this definition. Mindfulness is about focusing one's attention—intentionally. That focused attention occurs and is directed toward the individual's experience of the present moment, as perceived through all the individual's senses. Finally, the focusing of attention is non-judgmental. Mindfulness is about noticing what is there without engaging in other mental processes, such as evaluating, sorting, or labeling. As you focus your attention, at some point, you notice that thoughts have emerged and pulled you away from what you were focusing on, shifting your focus. When that happens, the important thing is to notice, and then intentionally turn your attention back to the object of focus. Likely, this will be something you have to do repeatedly. Losing your focus is not a failure—but a natural process. Please, take a few minutes to try out a mindfulness exercise adapted from one written by Dr. Robyn Walser and Dr. Darrah Westrup.[12]

* * *

EXERCISE 2.4 Mindful Breathing

I invite you to begin this exercise by placing your feet firmly on the floor and sitting upright so that your back is straight but relaxed. If you wish, close your eyes to limit distraction. Now, focus your attention on the end of your nose and breathe slowly. Notice the feeling in your nostrils of air moving in and out. You might notice that the air coming in is cooler than the air moving out of your nose. Simply let yourself gently observe your breath, paying attention to the air as it flows in and out. You might also observe sensations in your chest as it expands and contracts. If you become distracted by thoughts at some point, just take a moment to notice the content of the thoughts, where your mind took you, and then, without evaluating, release the thought and return your focus to your breathing. If you get distracted many times, return to noticing your breath each time. Simply continue to focus your attention on your breath for the next several minutes.

When you've finished, pause and reflect on the experience for a few moments. What did you notice? What sort of thoughts did your mind put forward? Were they thoughts about things you needed to do? Were there thoughts evaluating what you were doing? If so, remember, this is natural. The work is to return your attention back to whatever you have focused on—in the example above — the breath. This will help you remain aware in the present moment, able to choose to focus your attention purposefully. Strengthening this ability makes us more able to choose actions that are consistent with our most important values.

* * *

Practicing mindfulness regularly strengthens a person's ability to focus intentionally on the present moment's sensory experience as delivered to us through our five senses. It also builds the skill of noticing the constant and ever-changing content of our thoughts as an observer. This ability to notice our bodily experience in the moment, and to observe our thoughts and feelings, is one primary element of the practice of awareness.

As FSHD progresses and mobility becomes increasingly challenging, another form of mindful awareness becomes necessary. For me, walking mindfully has become a necessity if I need to keep from falling and injuring myself. There's no longer any room for walking while on autopilot, casually looking around at the landscape, or fully engaged in a conversation with another person walking beside me. If I don't focus single-mindedly on putting one foot in front of the other without stumbling, bad things can and have happened. All that being said, I also find that I can get so wrapped up in my struggle to keep from falling that I miss what's present in the here and now. I find it important during the day, especially when I'm outdoors, to find a safe place to sit down or to stand still, and then to use those mindfulness skills observing and appreciating what's all around me by paying attention to each of my senses.

Self as Context

The second process that is a part of the practice of awareness is called "self as context." What do we mean by this term? Self as context stands in contrast to another term, "self as content." The way we use language is an important part of understanding the difference between these two terms. When I say or think things like "*I'm angry*" or "*I'm stupid*," the language that I use conveys that I AM the content of my experience. The language equates the self with the emotion of anger or with the descriptor of stupid. Phrased in this way, the self becomes the emotion or thought. Self as context helps an individual become more aware of a sense of self that is separate from the content of their experience, whether it be emotions, thoughts, sensations, or memories.

Instead, the "self" is an enduring place or "context" where momentary experiences such as angry emotion or thoughts of being stupid are experienced. Self as context exercises allows us to notice the observer self, simply observing the ebb and flow of inner experience without being controlled by it. Self as context, provides the space for emotions and thoughts to rise and fall. From this perspective, negative emotions and thoughts are no longer threatening. They are not the self. They are not you. Instead, you are the place where they occur. When you can observe this quality of experiencing, you create the opportunity to choose freely how to respond to those observations. In this way, we can choose actions that move our feet in the directions that matter most to us—toward our values (no matter the internal experiences).

EXERCISE 2.5 The Observing Self

For the next minute, sit quietly and listen to the thoughts in your mind. Depending on the moment-by-moment pace of your thoughts, they may come slowly or in a rush. Simply notice them. Take your time, don't rush.

So there it is! That's the whole exercise. You just showed that there's a part of you (your mind) that speaks - that is your thinking mind AND there is a part of you that stands back away from the content of your mind and simply listens and observes — that is your observing self. Your observing self is that place where thinking occurs.

Let's do this exercise again. This time, sit quietly and focus on your sensations–noticing what you are feeling in your body and on your skin. If thoughts or evaluations about the sensations pop up, simply notice the thoughts and refocus your attention on what you are feeling in your body. Again, take your time, noticing sensations throughout your body.

Again, we see there's a part of you that experiences sensation, and there's also that the part of you that can simply observe those sensations. Your observing self is that same space, the same context where thinking and sensations occur.

* * *

The observer part of you is your self as context. Suppose you are the context for the content of your life (thoughts, sensations, emotions). In that case, you are not the content itself, then there is the space and freedom to choose, to respond intentionally. If we get too wrapped up in our thinking self and are only aware of the content of our mind and feelings, we risk giving up our freedom to make choices based on our values. If we hold too tightly to those thoughts and emotions, we may constrict our available choices.

Not only is our self as context the space within us where we experience thoughts and sensations, our self as context also provides the space where we can freely and intentionally choose what we will do. When we get too caught up by the content of our experience and cannot notice the part of us that is separate from that content, we may also fail to notice the space that is available for us to make alternative choices at that very moment.

A straightforward way to practice this sort of awareness is to notice and change the language you use. When you notice using language that equates the self with the experience such as "I'm so stupid" you can notice that expression and intentionally change the words you use. You might say instead - "I'm having the thought that I'm stupid." or "I'm having the emotion of anger." While your mind might tell you not to bother with such language changes, you may find that simple changes in language help bring into your awareness the presence of a you that's separate from the content of your mental experience.

Why would that matter for someone with FSHD? I find life brings constant reminders of the limitations associated with my disease. When reminders of lost abilities come to mind, they often elicit mental evaluations of my current status. Those evaluations may suggest that it might be just better to give up and hide myself away so I don't come into contact with those reminders anymore. I find it important to notice these thoughts as simply thoughts, and also to observe that I can stand apart from those thoughts, and make intentional choices about how I want to live and be in the world. I can choose not to give up, and to not isolate. Even in the presence of thoughts telling me I should go away and hide, I can choose to continue to engage in activities that matter to me and interact with people in the world.

My family has several photo albums, some that my parents put together and others that my wife and I have collected. In one, there is a picture of me at around age 6 standing on the porch in a suit and tie—dressed up and ready to go to church on Easter Sunday. There's another picture of me a couple of years later in a striped t-shirt proudly standing over my bicycle, ready to ride away. In another there's a photo of me in college with much longer hair sitting in a chair playing my guitar. Finally, there's a photo of me (cover of this book) in my 60s walking in the sunshine.

The faces in each of those photos look entirely different from one another. While an observer looking carefully at the photos might determine that this is the same individual at varying points across a lifetime, a casual observer might equally think these are different people. As I look at the photos, I realize that I've seen each of these faces looking back at me in the mirror at various times in my life, but I also recognize that it's the same ME in each of them. There's a continuity. While each of those selves saw the world and understood life a bit differently than now, I still contain all of that and more.

Think back to your own experience across time. You probably have your own photographs of yourself in different time periods. You certainly have your memory, and can think back to see the you that existed in that time. Reflect on each of those people who were you. What did they look like? How did they walk and move? What did they like to do? Who did they like to be with? What did they care about passionately? Your FSHD may have changed the way you experience life now, from the way you did at some earlier point. The important thing to notice is the continuity across time and across all the physical changes. You are still you! And no matter what the future might bring in terms of physical changes related to your disease, you will still be you.

As we live our lives, our minds weave our memories together into the story of who we are. This story includes the thoughts we have about ourselves and the labels and evaluations we use. We weave each into this narrative. Some might call this our identity. This is a natural and normal thing that people do. It's sort of shorthand—a summary of ourselves. All of that is well and good as long as it’s flexible - as long as it works and provides a meaningful way of being in the world and living life. At one time in my life, if you asked me to describe myself, I might've said, ”I'm a musician, I play classical guitar.”

Classical Guitar

At a later point in my life, if you'd asked me, I would've said, “I'm a clinical psychologist–I help veterans with PTSD.” If you asked me right now, I might say,” I'm a man with FSHD – my wife and I lead the Bay Area chapter of the FSHD Society.” Each of those identities was true, but none of these were the complete picture. I was and still am more than a psychologist; I was and still am more than a guitar player; I was and am still more than my FSHD… and so are you. I still love and listen to classical guitar music, even though my hands no longer function well enough to play it. My guitar still sits safely in its case inside my house. I’m still a licensed psychologist and approach varying aspects of my life from that perspective. I'm still involved in psychological consultation, research and writing. I'm also a person with the disease FSHD, a disease that places increasing limitations on my mobility, and makes it difficult to do certain things that used to be easy. I will always be more than my FSHD, no matter how much mobility I lose…. and so will you.

The practice of awareness that makes up the second of the three legs of the ACT stool increases our psychological flexibility. Holding too rigidly to mind-generated conceptions of who we are is limiting and imprisoning. Awareness can help us see our freedom to live in the present moment in a way that is consistent with the things that matter most to us.

For me as a person with FSHD (and perhaps for you as well), I find it easy to get caught up in and focused on my personal physical struggles, to where I often miss precious experiences and people that are right there in front of me. Developing the ability to be present, and experiencing a sense of myself that is bigger than my disease, has improved my day-to-day life experience. Because this disease is chronic and progresses inexorably over time, it is easy to get lost in thoughts about the way things were, or in worries about what the future holds. Practicing awareness can bring our attention back to the present moment, where we can make choices and take actions that improve our life in the here and now.

Doing what matters

The last leg of the three-legged stool of ACT is the practice of doing what matters. This practice, as with the others, has two distinct core processes. These processes help me live a more meaningful life engaged in activities that truly matter to me. While that statement is true, I have also found that it’s not so simple, at least for me. I consider myself lucky to have had a career focused around the expression of one of my important values - helping others. However, trying to balance work with competing values of being a loving, consistent part of a family, and maintaining my physical health, has often been a challenge, especially toward the end of my career. Life has a way of throwing a wrench into the works, and disrupting our careful balance with difficult, unexpected challenges. The death of friends or family, financial chaos, disease, accidents or injury - these experiences don’t ask permission before disrupting our lives. In 2018, I retired from my work at the VA, and immediately realized I now had to radically re-orient how my I lived my life and how my I expressed my core values. Five years have passed since retirement and figuring out who I am now and what's important to me now is still underway. Life during the COVID-19 pandemic introduced new and painful challenges for all of us. Even at age 68, I remain a work in progress. Making these core ACT processes a part of everyday life is a key part of becoming more psychologically flexible. Figuring out how to continue to do what matters has been a big part of my transition into retirement. I genuinely believe that it will be an important part of your journey toward a more meaningful life as well.

Values

The first of the two processes that help us engage in the practice of doing what matters is values. Values have a particular meaning within ACT that sets them apart. Years ago, as part of a psychoeducational group that I led, I used to do an exercise with group members. I would hand them a blank piece of paper. On one side, I would ask them to think for a moment and then write their most important value. Often it would be something like family or courage or something similar. After they finished, I would have them turn the paper over and draw out a calendar week with each of the days marked. I would have them imagine a typical week and write for each day of the week how they would typically spend their time during the morning, afternoon, and evening. Then I asked them to reflect on how they spent their time, determine what activity occupied most of the time, and suggested THAT might be their lived value.

I particularly remember one veteran who had written "family" on the first side of the paper. Leaving aside for the moment the fact that family is not a value - rather, family is a set of relationships where we live and express values. On the other side of his paper, he described spending most of his day and evening shut away in his room with the door closed - drinking, and watching TV. He realized perhaps he was valuing isolation as opposed to being a loving family member.

The question that arose for that veteran is also a question for each of us. What does it mean for us if the thing that we value most is not the thing that we spend most of our time actually doing?

From an ACT perspective, values live in our feet. Values are not simply mental ideas or beliefs. Rather, values can only exist as they come to life through our actions. Values only have meaning; they only become real when brought to life through actions in the present moment. Values are not goals. They cannot be identified, performed, and checked off a list as complete—like items on our to do list. Instead, values are more like the directions on a compass. A magnetic compass points you north. By looking at the compass needle, we can always find a path to continue moving in our desired direction.

Compass

A compass can guide our movement, but it actually doesn't move our feet. In this way, values serve as a guide for our actions. I live right now on the West Coast. Suppose I were to take a compass and decide to travel east. In that case, I could begin traveling in an easterly direction forever with the right vehicle. I could cross the US, then the Atlantic ocean, Europe, Asia, then back around over the Pacific ocean and continue if I wanted. All that time, I would travel east. Values are similar in that I can continue to perform actions consistent with my values without ever running out of things to do. Values can be our guide to help us choose the actions that will lead in the direction that brings meaningfulness, purpose, and joy to self and others. However, amid all of that movement toward values, we will also carry with us and continue to experience all the other parts of life, including sadness, loss, and various sources of pain, both physical and mental, that come with FSHD.

One element of the practice of doing what matters involves identifying our true values. The following is another exercise adapted from one created by Dr. Walser and Dr. Westrup.[11] Please take a few moments to read and engage in this exercise carefully to identify your values.

* * *

EXERCISE 2.6 Lost Values

Instructions: For this exercise, find a quiet place without disturbances, and where you have a place for writing. Bring a pen and some paper with you to have alongside. You will need them as the exercise proceeds. We will begin this exercise with mindfulness. As we have in previous exercises, nestle into a chair. Place your feet firmly on the floor. Let your hands relax in your lap or on the arms of the chair. If you are comfortable doing so, close your eyes so you can pay attention to your breathing.

Begin breathing in and out slowly. Simply breathe in, and then breathe out, slowly… comfortably. Focus your attention on your breath and notice the sensations within your body as you breathe slowly in - and then out. Notice how the air feels as it passes into and then out of your nostrils. Notice the difference in temperature as the air first passes into your body and then notice the warmth as it returns from your lungs and out through your nose or mouth. Take a moment to notice the sensations in your abdomen and chest as air inflates your lungs, and then compresses to push the air back out. Simply breathe, and allow yourself to notice your breathing for about two minutes.

After a couple of minutes, reflect on your life history. Think back across your life span (childhood, adolescence, early adulthood). Think about what was most important to you during each of those times. How did you typically spend your day? Who were your closest friends? What were the things you did that truly mattered to you in those years? Continue to reflect, noticing the values you had then, and importantly, notice any values that may have gotten lost over time or pushed away because of the progress of your FSHD.

What are the things that you've stopped doing that used to matter to you? Notice any costs associated with the losses of those values, but be careful to remain nonjudgmental. Take some time to reflect honestly on the things that mattered during each of these periods of your life. What changes in what you do and what matters to you has your experience of FSHD brought? What changes in activity and values have come about simply through growing older and maturing?

After this review, gently bring yourself back to the room. Now try to imagine that you could breathe life back into these lost values. Imagine that the gift of vitality has returned to these values. Write a paragraph about each of these lost values and the importance they had for you then. Be kind to yourself as you do this exercise. Notice any self-critical thoughts that arise, then return to the exercise. After you finish writing, read through what you've written and circle each value that you find.

Finally, make a list of several things for each value you have circled that you could do within the next twenty-four hours to bring these values back to life. Challenge yourself to engage one of these activities in the next 24 hours.

* * *

The last part of the exercise above introduces us to our final ACT process - taking action. Taking action pulls together all of our efforts to engage differently with our disease - with FSHD. Taking action is an important element of learning to create meaning in the face of FSHD. I have found that by choosing to take action in several areas of my life, I've made my experience of living with this disease more manageable, and my life more meaningful.

Committed Action

The second process, together with values, that comprises doing what matters is what is called committed action. One way to define committed action is to think about what it is not. Committed action is not impulsive or reactive. Committed action is not an action taken to avoid some unpleasant internal experience. Instead, committed action is a conscious, intentional choice to take a values-based action - right now, in the present moment. This means behaving in ways that are consistent with what you care most about. Sounds like it should be easy, right? Once you know what's important to you, you just do it! Simple. Why I am I telling you this?

If committed action were as simple as it sounds, we would all be doing it already, and I wouldn't be writing this book. The seeming internal barriers associated with FSHD become barriers to action. I am writing this book to let you know that this doesn't need to be the case. You can have FSHD and still live and act in ways that are linked to your values. You don't have to feel and think differently before you can live well, even with FSHD.

I have fantastic news for you, along with tough news. The fantastic news is that every human being has the power within them in every single moment to choose a direction and take action. Every single new moment is a unique opportunity to live the life we truly want. Now, here's the hard news. It's not enough to just turn in the right direction. Committed action is not a one-and-done experience. It requires sustained, persistent movement. It's taking a step forward, and then another, and another, and another, without stopping. This part requires all three legs of the ACT stool to work together to support us.

We live in a world where the expectation is instant gratification. We want what we want when we want it. Our TVs and streaming services have thousands of channels and options, so that what we want is instantly available. We shop in megastores or on Amazon so we can immediately find exactly what we want and buy it. If it's not right there in front of us, we can have it shipped for free and delivered in just one day. We multitask and rush from task to task.

All of that is out of step with committed action. Committed action means sustaining our focus in a values-based direction and continuing to move in that direction, one action at a time, moment-to-moment, day-after-day, week-after-week, month-after-month, year-after-year. Committed action requires a willingness to persist even when our emotions and thoughts tell us to stop or that we can't continue. It requires noticing when our attention waivers, when we head in a different direction, or noticing that we've stopped all together. At that point, committed action is about choosing to re-orient ourselves and start again.

It occurs to me, that I'm using language about taking steps, walking, and moving about–when I am writing to people who like me have FSHD, and actually have a great deal of physical difficulty with things like walking. FSHD interferes with my mobility, and probably yours too. When I talk about taking actions that are consistent with my values, those actions may or may not involve physical movement or walking. I may need to roll my wheelchair toward my values. I've learned that I can continue to live in a way that expresses my most important values even when FSHD impairs my physical ability to move. Let me give you an example.

One thing my wife and I used to do together with friends was go to local street fairs. We would enjoy walking around, looking at various booths where artists or craftspeople displayed and sold their goods. We enjoyed spending time with and talking to our friends. As my mobility difficulties grew, standing and walking for a couple of hours became increasingly difficult. I would be constantly looking around for a place to sit, and thinking about how I would like to leave and go back to the car. Eventually, when we got invited to meet friends at such an event, I would say I didn't feel up to it. The reality was I was resisting using mobility aids such as AFOs, or a walker, or wheelchair. The prospect of being seen using those aids and being perceived (or perceiving myself) as disabled amid a crowd of people brought up uncomfortable emotions that I wanted to avoid. But that avoidance was threatening to cut me off from friendships and support.

Finally, I realized that if I were going to take actions consistent with the value of friendship and being a good friend, I would need to accept having the unpleasant emotions and thoughts that might come with using mobility aids in public. Over time, what I discovered is that those thoughts and emotions subsided, and that being willing to have those emotions, opened up space to have other experiences and other emotions such as the joy of being with friends again, and seeing and doing new things. Engaging the ACT processes of willingness and defusion helped me to re-engage in committed actions directed toward strengthening my friendships. I have benefited from engaging in committed actions consistent with my values. Perhaps you will too.

At the start of every new year, millions of people make resolutions to change. Whether it’s losing weight or beginning an exercise program, millions of people decide to change something in their lives. Most don’t sustain those changes. Instead, in a few weeks or months, things have returned to the way they were. The resolution no longer holds its resolve.

In physics, Isaac Newton's first law of motion is called inertia. That law states that objects keep on doing what they’re doing and resist changes in their state of movement. That tendency is true for people too, but it’s both bad news and good news. Making changes takes intense effort and energy to overcome the inertia of our existing habits and patterns that allow us to live and function mostly on autopilot. However, once you make and sustain those changes, the good news is that they also get locked in, protected by that same inertia.

So what happens with the New Year’s resolutions? Why do people set out towards making important life changes and then cannot continue? Likely, there are many reasons. Observable changes may not have come quickly or easily enough, and they stopped trying. More than likely, everyday stresses (including other competing values and other important activities) presented obstacles. This likely sapped their motivation and made following through with their new commitment increasingly tricky.

When deciding to engage in the course of committed action focused on pursuing a core life value, it is essential to reflect on obstacles that are likely to emerge. For instance, if I were to make a New Year’s resolution to improve my physical health through exercise, it would be important to reflect on why I am not exercising regularly now. Are there obstacles in my living environment that may make exercise more difficult? These could be things like wanting to go jogging but not having running shoes or wanting to work out at a gym but not having a gym membership. Scheduling could also be part of the difficulty. Perhaps, I work long hours and don't get home until late. I feel too tired to work out in the evening. Perhaps, I also stay up too late and have difficulty getting up early to exercise before work. With any of these barriers, a certain amount of problem-solving will be required to overcome the challenges. This is an excellent place to engage my mind. It can help me problem solve.

However, there may also be other kinds of obstacles where engaging my mind may not be useful. Specific experiences will arise from inside my body. Suppose I am overweight and feel embarrassed. In that case, I might want to avoid being seen wearing exercise gear outdoors or at the gym. Maybe my mind tells me things like *"you can't do this, you won't succeed,"* or *"you've tried before, and it didn't work."* When internal obstacles such as painful thoughts and emotions arise, the ACT processes we've explored, such as willingness or defusion, will be necessary to follow through on my commitment. I will need to disentangle from my mind and be open to whatever emotions arise, no longer letting them choose for me. Instead, choosing to engage, following through on what I value.

Look at your own experience. What are the obstacles currently preventing you from taking committed actions that express your deepest values? What are the obstacles that FSHD presents? Where do you think you might get tangled with thoughts and emotions when you are trying to take committed action? Try defusion and see what happens.

Another vital element of committed action is paying attention to potential blind spots. What do I mean by that? Let's say that it's true that I work a lot of hours, and that my job is important along with my family, and I've committed to improving my health through exercise. Perhaps, my partner is missing me because of my long hours at work. She has complained about my not being around in the evening to assist with chores and parenting. If I were to decide to begin a routine of going to the gym after work, I could view it as a committed action to pursue the value of health.

However, going to the gym after work could also function to avoid emotionally unpleasant conversations with family members about the hours spent away from them. My committed action in pursuing health may actually function to avoid unpleasant emotions resulting from discussions related to another value—being a loving and engaged family member. Noticing how a particular action functions in my life is key to determining whether it is truly a committed action or simply a way of avoiding unpleasant emotions, and ends up functioning as an obstacle to another important value. Ask yourself - is my action functioning to avoid unpleasant emotions, thoughts, or memories?.

Most of us have more than a single important value, we hold multiple values. Living a meaningful life and pursuing multiple (sometimes competing) values can be challenging. It's necessary to balance, prioritize, and make discriminating choices about the specific actions we take in pursuit of our values.

It’s important to remember that committed action is a process. It’s not something that’s done once and is complete. Rather, it’s something that we continue to choose day after day, and if we notice that we’ve gotten off course, or even stopped, in that moment we can again choose to get moving. The commitment is to continue. Just as FSHD is a disease that continues progressing, and never really stops - it may slow down or speed up, but never goes away until there is an effective treatment or cure. In the same way, we can choose to engage in these ACT processes, sometimes falling down, sometimes speeding up, paying attention, creating space to make choices that move us closer to our values, and then choosing to take action. The following exercise is a single step in that direction, one that you can use repeatedly in the future.

* * *

EXERCISE 2.7 Bold Move

In the table below, I will ask you to identify an important value, perhaps one held in the past, that may have fallen by the wayside. Take a moment and write one or more of those values into the lines in the left-hand column.

In the second column, I invite you to list several tangible actions you could take today that would give expression to that value. It could be anything, big or small, but it should be something you could act on today, right now, while you pause from reading this book.

Finally, in the third column, I want you to list what has prevented you from taking action until now. Think a little more about what might get in your way. Are there additional steps that you must accomplish first? Are there thoughts or emotions or memories coming up that are unpleasant? Write those things down in column three.

Now, for the most crucial part of this exercise. Pick one of those actions you listed in column 2 and do it–right now. If you have questions about whether it's appropriate in some way - pick a *different one. You can always consult with someone and check the other one out later. Find one that is doable now, and take action. Return to the book when you have finished.*

<table>
<tr><th colspan="3">Bold Move List</th></tr>
<tr><th>Value</th><th>Actions I can take that express that value</th><th>What obstacles might get in the way?</th></tr>
<tr><td></td><td></td><td></td></tr>
<tr><td></td><td></td><td></td></tr>
<tr><th colspan="3">Observations</th></tr>
<tr><td colspan="3"></td></tr>
</table>

Take some time to reflect on the action that you took. What came up? What thoughts and feelings did you notice before, during, or after taking action? Take a few moments and jot down some notes about what you noticed in the box titled "Observations." Remember! Values are like a compass direction. You can continue your journey in that valued direction by identifying and taking more actions now and in the future.

* * *

In Part Two, I've provided an overview and rationale for Acceptance and Commitment Therapy (ACT) especially as it applies to FSHD. There are many books already written on ACT related to other common issues and problems, so this section is just a brief introduction. I see this section as a quick overview and guide to ACT, that you can come back to again and again if things get confusing as you move forward in the rest of the book.

I'd like to summarize what we've covered so far, so that you can keep it in the front of your mind as you continue to read the rest of the book. We began the section talking about the universality of human suffering. When I was in my 20s, I felt like I was at the top of my game. My mind told me I knew everything I needed to know about how to succeed in life. My mind was wrong! I've discovered that life is so much more difficult than I ever imagined as a younger man. Life brings difficult, unfair, immense challenges, often beyond what we believe we can bear. Aside from the bodily and environmental challenges and difficulties that we have to cope with in the physical world, our mind creates additional mental suffering. We often try to deal with internal mental challenges in the same ways that work so well in the physical world - we try to avoid or control them, but often discover that our efforts are unsuccessful, or that they even increase our suffering.

ACT proposes that the way forward to a more meaningful life is through building psychological flexibility. We defined psychological flexibility using Dr. Darrah Westrup's definition as: the "*ability to respond to life in a workable way, a way that enables us to live vital, meaningful lives.*"[3]

We described ACT as comprising three practices (Opening Up, Awareness, and Doing What Matters) using the metaphor of a three-Legged stool. ACT uses this metaphor to suggest that each of these practices is essential to support psychological flexibility in the face of life's challenges.

Similarly, each of these practices has two core processes associated with it. Acceptance, and Defusion are the processes that make up the practice of Opening Up.

We can think of Acceptance as a continual process of meeting our internal thoughts and emotions with nonjudgmental openness, and a willingness to be present with whatever painful or pleasant experience arises within us - without responding with attempts to avoid or control them.

Defusion is the second process involved in Opening Up. You can think of defusion as loosening the grip that our thoughts can hold on our behavior. Diffusion is the skill of learning to see our thoughts, simply as thoughts, taking away their power to control our behavior. You can also think of defusion as the ability to step back from our thoughts and to open up space to make intentional choices about the things that we hold to be important and in the actions that we will take as we live day to day.

The practice of Awareness also has two distinct processes. The first of these is Present Moment Awareness. In reality, the present moment is the only place where life is lived. Though we can use our present moments to ruminate about past events, or to worry about future ones, the present moment is the only place where we can actually live out the things important to us. Mindfulness is a skill that can expand our ability to be aware and engaged in the present moment. We introduced mindfulness in this section, but recommend that you each devote time to practicing mindfulness - using exercises you'll find in the rest of this book, as well as in many other places.

The second process of Awareness is called Self-as-Context. The process of self as context makes us more aware of our observer self. It helps us to create a sense of distance from the “content” of our inner experience, whether these are emotions, thoughts, sensations, or memories. Instead, the self becomes the place where we experience momentary thoughts, memories, sensations, or emotions, while maintaining an awareness of one’s self that is larger than and extends beyond those experiences. Self as context exercises allow us to become more aware of the observer self, able to notice the ebb and flow of inner experience without being controlled by it.

The third leg of the stool - the practice of Doing What Matters also has two processes. The first of these is Values. This process entails identifying those things that are most important, as well as what actions we can take in the present moment that will move us closer to those values. Values function like a compass - they guide our way, keep us on track, and help us find our way back to our chosen path when we get stuck or lost. Values, however, do not move our feet. They may motivate us, but a second process is required for movement to occur.

That second process is called Committed Action. Committed action is where the rubber meets the road. Committed action is the culmination of all the ACT practices and processes. If our lived experience with FSHD (as well as with the many other challenges that life brings) is to change, and the meaningfulness of our lives is to improve - committed action (along with the other processes) will be at the center of the change. Committed action requires persistence and dedicated effort, but I’ve found that effort to be worthwhile.

I hope this book will be a useful tool for you as you confront the day-to-day life challenges of living with FSHD. In Part One of the book, I summarized FSHD and the research evidence that suggests there is a need for mental health resources in managing the disease. In Part Two, as I’ve mentioned, I’ve outlined the core principles of Acceptance and Commitment Therapy (ACT) to help you understand where we’re going.

The third and final section of this book (Part Three) will flesh out in much greater detail the ACT processes as they specifically relate to seven common themes that are relevant for people with FSHD. You should view part Three as a workbook. I would suggest that you start by reading it all the way through initially. But after that first reading is complete, I would suggest setting aside time, and focusing on a single module from Part Three and working through it carefully for an extended period - think days or weeks - not minutes or hours. Do some of the individual exercises daily for a period. Try to take enough time with each of these exercises to discover where they might be useful in your journey with FSHD from now on. When you feel you're finished with a module, pick another one and spend an extended amount of time working through it as well. Changing the automatic, habitual ways that we face the progressive and ever-changing experience of FSHD will require persistent effort, reflection, and time to accomplish and maintain. However, I truly believe that the rewards of doing this will far exceed the costs. Please know that my best wishes and hopes are with you as you undertake this challenge.

PART THREE

ACT self-help exercises to support those struggling with common FSHD experiences

"If you don't make time for exercise, you'll probably
have to make time for illness."

- Robin Sharma

In Part One, I summarized the physical and mental impact of FSHD, both from my personal perspective and from research. In Part Two, I briefly reviewed the rationale for ACT as a therapeutic approach and briefly explained the core practices and processes found in ACT. You can think of the first two parts of this book as education about FSHD and ACT. The sections have important information for FSHD patients dealing with the disease's effect on their emotions and daily life..

Part Three is where the rubber meets the road. In this section, we will apply the therapeutic processes of ACT to seven common themes that frequently occur in the lives of those with FSHD. These themes come from research, my FSHD experience, and conversations with others who have the disease. You should think of this third section as your personal workbook. It will require focused attention, energy, and time to benefit from the modules in this section. Indeed, initially, you'll want to read through each module and the exercises contained within this section. However, simply reading likely won't be enough. I hope you will spend time with each of these seven modules. Reflect on how you experience these themes within your own life. I suggest engaging in each of the exercises provided, and developing and practicing mindfulness.

Paying Attention and Acting are Power: Overcoming Avoidance

In Part Two, we discussed the problem of experiential avoidance and its role in making life increasingly inflexible. Inflexibility means we are liable to get stuck - and have difficulty continuing to move forward. Parts of our life become increasingly unworkable. We lose out on opportunities to pursue valued meaningful activities that would improve our quality of life. I have to confess that for many, many years, I took an avoidance approach to my FSHD.

When I first learned that my FSHD was lifelong, degenerative, and that there was no treatment, my attitude shifted. It made no sense to go to the doctor just to hear what I already knew, and to get no help. In the first 20 years after my diagnosis, I only had two appointments with a neurologist knowledgeable about FSHD. In both instances, I sought the appointment because of the emergence of some new physical manifestation of the disease. I first sought help related to the emergence of a mysterious pain. That visit clinically confirmed my diagnosis of FSHD. Eight weeks of physical therapy eliminated the pain and brought me back to baseline functioning. The second visit occurred at least 10 years later when I started stumbling and falling more frequently. That doctor made some specific recommendations that I ultimately ignored and failed to act on for another 7-8 years after that visit.

It took me years to realize that avoiding my emotions about FSHD made it difficult for me to come to terms with my disease. It also hindered my ability to experience a good deal of meaning and purpose, hope and joy. I now regret not taking advantage of mobility devices, physical activities, and other helpful supports earlier.

Avoiding the signs of physical decline caused me to overlook my overall health. This certainly occurred related to my FSHD, but also related to the development of other significant health conditions. It's emotionally challenging to measure and watch our health because it reminds us of the impact of this disease we didn’t ask for and can't control. By not addressing my emotional reactions to my disease, I also neglected other aspects of my physical well-being. I think that sometimes I've even punished my body by not taking care of it, even in ways that were self-destructive. I have to say clearly that even though staying away from my painful emotions about FSHD felt good in the moment; it created unhelpful consequences for my life quality.

In speaking with other people with this disease, I realized that I’m not alone in how I have responded. If you’ve read this far along in this book, you likely have FSHD yourself, or you know and care about someone with the disease. My hope for you as a reader is that you will fully use the tools and exercises that follow. Paying attention to my body, reengaging with the world, and with other people has made my life with this disease more meaningful. Learning to pay attention to how the disease is affecting me has required educating myself, and engaging in what I will call - self monitoring. I truly believe that practicing these things will be as helpful for you and your family as it has been for me and my family.

Self-Education about FSHD

As mentioned previously in Part One, research shows that understanding FSHD can help reduce illness perceptions associated with worse quality of life.[1-2] Learning about FSHD disrupts the tendency to avoid painful reminders of the disease. It brings us into contact with information about current research that is actually quite hopeful. It's challenging to gain access to the actual medical journals where researchers publish FSHD research. Without a strong medical research background, it can be hard for non-scientists to interpret these studies. To keep up with my FSHD, I check the FSHD Society and Muscular Dystrophy Association websites. I have found both these organizations critical in helping lead me to the information that I needed to improve my quality of life.

FSHD Society

Patients formed the FSHD Society in 1991. It is now the largest network of individuals with FSHD. Community donations of $10 million+ have funded FSHD genetic research, resulting in biotech companies expressing interest in treatment development. The current goal of the FSHD society is to have an effective treatment available to patients by 2025, blazing a path toward an eventual cure. There are many regional chapters throughout the US. The chapters meet regularly for education, support, and fundraising to advance research. You can find out more about the local FSHD Society chapters here. Indeed, my wife and I are the chapter directors for the SF Bay Area FSHD society chapter. Get updates on research and society activities through the FSHD Society's newsletter, the FSHD Advocate.

Muscular Dystrophy Association(MDA)

The MDA was created in 1950 to raise awareness about muscular dystrophy and to raise funds towards a cure. The organization funds research grants and care centers throughout the country. From 1951 through 2015, the MDA telethon was the primary annual fundraising event, hosted by Jerry Lewis for most of those years. The organization deals with many neuromuscular diseases, such as muscular dystrophy, but has a dedicated FSHD page. The MDA provides a downloadable fact sheet about FSHD here.

Self-Monitoring of FSHD and Overall Health

Self-monitoring is when you *intentionally observe and record important aspects of your life for later review.* The APA Dictionary of Psychology defines it as "*a method in behavioral management in which individuals keep a record of their behavior (e.g., time spent, form and place of occurrence, feelings during performance), especially in connection with efforts to change or control the self.*" It's just in the last 6-8 years that I have appreciated the importance of self-monitoring in my life. As I have aged, the pace of my FSHD-disease progress has increased, and my physical strength has decreased significantly. It has become crucial to monitor my FSHD health status and my health-related behaviors. Self-monitoring helps me notice small physical and mental changes. Self-monitoring is important for conscious and intentional living, even though it requires more effort than living on autopilot. It allows me to make smarter decisions and to direct my actions in ways consistent with my most important values. I believe that self-monitoring can help you in similar ways.

Creating a Journal

My 35-year-old self would be stunned to hear my 69-year-old self telling you this, but one way I have found useful in recent years to self-monitor is by keeping a very simple journal. My younger self would've pooh-poohed the idea of keeping a journal. Not that people hadn't recommended it to me, but I never thought that I would have anything worth writing about. The type of journal I'm talking about isn't really a diary or an autobiography. I don't keep a continuous narrative of each day or my life history, and it's certainly not something I write daily. There may be short periods when I jot something down several days in a row, but mostly it's every week or two.

I use journaling to pay attention to important things going on inside me and around me, especially as it relates to my FSHD. I might make a note about stumbling and falling and what was happening at that moment. I might write about spilling food on myself at a restaurant dinner, the thoughts, emotions, and self-evaluations that emerged inside of me. I write notes about things that trigger emotions related to FSHD experiences or losses. My journal is an electronic file that I keep on my computer. Whenever I have something to write, I open it. I put a day/date in every entry. Occasionally, I go back and read through parts of it. I find it most beneficial when I see patterns emerging in the experiences I have and the changes that are slowly occurring slowly. My journal provides a way of gaining insight into myself. It's a way of paying attention to my life, and to the progress of my disease. It helps me notice how FSHD affects other important areas of my life. I would highly recommend that you consider creating your own journal and tracking what happens in and around you, too. Paying closer attention can help you notice some costs that arise from avoiding dealing with the impact of your disease by living only on autopilot.

Self monitoring is not something we do for its own sake. Paying attention to our thoughts and physical changes is only helpful when it leads us closer to our values. Writing in a journal can be one way of doing that. Using more formalized measures to monitor changes is another way of paying attention.

Standardized assessments for monitoring health

Self-monitoring using questionnaires or physical tasks can also help you notice important changes. Health professionals use questionnaires and tasks in this section to monitor health. Using these tools at home from time to time keeps you aware of what's happening within you. Using standardized measures can also allow you to share helpful information with your medical team.

Disability Index:

Appendix 1 contains one straightforward questionnaire you might use. The Stanford **HAQ-8** is a brief screening measure that helps quantify a person's current level of disability.[3] There is also a 20 item version of the scale that might be slightly more sensitive to subtle changes in functioning. The score on the HAQ-8 scale is useful for quantifying one's overall level of disability. However, because it is brief and because FSHD is a slowly progressive disease, your score is unlikely to change quickly. Taking the instrument again every six months or a year can help you monitor how your FSHD is progressing.

Standardized physical tasks:

Standardized physical tasks are another way of monitoring one's physical abilities, particularly mobility. You can do some of these tasks at home as a way of monitoring your physical status. The **Timed up-and-go** task is easy (or maybe not) and quick to perform. Doing it regularly (e.g. monthly) can help you notice physical decline, stabilization, or improvement if you become more active.[4] You can do this task anywhere there is a chair and a flat open area free of obstacles.

Prepare for the TUG task by placing a sturdy chair on a stable flat surface where it will not slip or slide. With a measuring tape, mark a spot exactly 10-feet in front of the chair. If you are unstable on your feet, have someone else there to observe and time you.

If you need an assistive device such as braces, a cane, or a walker to walk, you may use them for the task. When you are ready to begin the task—sit down in the chair with your back fully touching the back of the chair, and place your hands comfortably in your lap. Using the stopwatch function on a phone, begin the timer. Now, stand and as quickly as you can - walk to the 10-foot marker, turn around, walk back to the chair, and sit down. Stop the timer and record your time. Do this task 2 to 3 times and take an average of the times. Record your time and the date in your journal and then retest yourself in a month or two. Video recording can give visual information about changes in your standing and walking..

The Six-minute Walk Test (6MWT) is a more difficult physical function measurement task.[5] The standard clinical procedure requires walking between two cones 10 meters away from each other. They instruct you to walk as fast and far as safely possible in six minutes. Total the number of laps completed and add the distance of the last partial lap to arrive at the total number of meters walked. You can do this task at home walking around a small residential block or up-and-down the street. Several smartphone apps designed for walking, running, or cycling can calculate the total distance traveled using GPS. These apps may not be accurate for short distances and GPS signal variation can make it difficult to track minor changes in walking ability. To get an accurate measurement, buy children's sidewalk chalk and a measuring wheel for a few dollars. Then, do the six-minute walk in your neighborhood. Measuring wheels are available for less than $20, and can accurately measure distances up to 10,000 feet - which is further than any of us can walk in six minutes.

We've talked about monitoring physical functioning. Now we’ll turn to ways of monitoring your psychological functioning.

Measuring Depression

I have noted previously that many individuals with FSHD report at least some symptoms consistent with depression. The Patient health questionnaire (PHQ-9) is a very brief screening measure for depression.[6] You can find it in **Appendix 2**. The scale is easy to administer and score, with a total score showing the level of depression present. PHQ-9 scores of between 5-9 suggest mild depression, 10-14 suggests moderate depression, 15-19 suggests moderately severe depression, and 20+ suggests a severe level of depression.

Measuring Psychological Flexibility:

Suppose you're continuing to work through these varying exercises designed to improve your psychological flexibility, and hopefully, the quality of your life. In that case, it will be useful to monitor your progress. Research often measures psychological flexibility before and after working on the ACT exercises. Included in **Appendices 3 & 4** are two questionnaires that measure elements of psychological flexibility[7-8]. If you're ready to work through the seven modules that follow, I would strongly encourage you to complete both these questionnaires before you begin. This will reveal your current level of psychological flexibility. Complete the questionnaires and follow the instructions for scoring. Save your scores for each questionnaire - it will be useful to compare your scores after you work through the seven modules. Record the scores that you receive and then re-take the questionnaires when you’ve completed working through all the material in the book. Using these questionnaires can help you see tangibly whether you are receiving a benefit from these materials and exercises. However, the genuine test will be the quality of your life in the future.

After you have completed the questionnaires, I encourage you to work through the seven ACT for FSHD modules that follow. Over the next few weeks or even months, take your time working through each of the modules. Each module focusses on a specific theme, which may or may not be entirely relevant to your unique FSHD situation. I would encourage you to give each exercise a try, regardless. ACT is not something easily explained or understood. It's only by experientially doing these exercises that one really begins to “get" how ACT works. Engaging in each of these modules and working through them could easily take a month or more. Don't rush! It is more important to work through them with care than to simply finish.

Merely reading a book does not produce personal change. Change requires focussed energy and actions taken consistently over time to see results. I would suggest that you plan to spend as much as a full week, or more, with each module working through the exercises. Each day, pick one exercise and work through it. Keep a pad of paper handy (or use your journal) to jot down notes about your observations. Write about your thoughts and feelings. Write specific actions that you intend to take. Set aside time each day to return to the exercises. Do a mindfulness exercise daily from this book or other sources.

Keep in mind the overall goal for ACT and for this book is to live well. The goal is to help you become increasingly flexible. I'm not talking about physical flexibility here, though that also is a good thing for many of us with FSHD. I mean, becoming more internally flexible. Expand your life, take emotional risks to engage in meaningful activities that you've avoided out of fear or hopelessness. Internal flexibility is about finding and pursuing important values with effort and energy.

What follows in this section are seven modules. The topics for each of these modules are drawn from my personal experience with FSHD, from conversations that I've had with other individuals with the disease, and from the research literature about FSHD. Each module begins by laying out the overall topic of the module along with several goals that I hope you'll be able to accomplish. Each module provides several exercises that will allow you to begin to experience and utilize one or more of the six ACT core processes.

The first two modules focus on our internal experience of "thinking." I mean the times when we get stuck with thoughts that spin round and round repeatedly in our head. Sometimes we call episodes of continuous thinking about past events - rumination. We call it worrying when we direct repetitive thoughts towards future events that have not happened yet. Sometimes during periods of thinking like this - emotions of various types also show up - either before or after the thoughts occur. If the thoughts have a negative tone, often these emotions will feel painful or negative, too. Module 1 focuses on thoughts and emotions that show up related to memories and reminders of things FSHD has taken away from our lives. Module 2 focuses on thoughts and emotions that show up related to worry about things that haven't yet happened but might happen. Each of those modules provides exercises help us be more flexible in how we engage with those thoughts and emotions.

Module 3 focuses on "body image" for people who have FSHD. Our society pays huge attention to physical appearance. We learn extremely high and unrealistic expectations about physical appearance. As a result, many people in the US believe they cannot meet those societal expectations. Society stigmatizes those who don't meet physical expectations, particularly the visibly disabled. The exercises can help us become more flexible with the thoughts that show up about our physical abilities and appearance..

Modules 4 and 5 focus on two highly prevalent FSHD symptoms: physical pain and chronic fatigue. These two symptoms interact with physical activity in ways that make FSHD difficult to manage. These modules use exercises proven helpful in other patient populations that struggle with pain and fatigue.

Module 6 focuses on managing one's body weight when one has FSHD. Loss of muscle mass because of the disease can lower an individual's metabolic rate and lead to weight gain. Increasing weight and decreasing strength can put people with FSHD at risk of falls and physical injury. This module provides tools that have proven helpful in other populations that struggle to manage weight.

Module 7 extends the knowledge about ACT to the entire family. The physical changes FSHD brings can affect both the individual with the disease and the people that live with them. The financial burden on the family can affect spouses and siblings. Caregiving responsibilities may affect life choices for them too. The last module describes some family tensions and developmental challenges. It includes exercises to improve psychological flexibility within the family.

When you have completed all the modules that you choose to complete, go back to the questionnaires listed above and retake them. Compare your scores to see if they show improvements in your psychological flexibility. Note, the ultimate test is not your numeric score. It is whether you are engaged in living out your values and moving forward toward the things that matter most to you. That's my genuine hope for you. Good luck on your journey! We now turn to the workbook portion of this book.

MODULE ONE

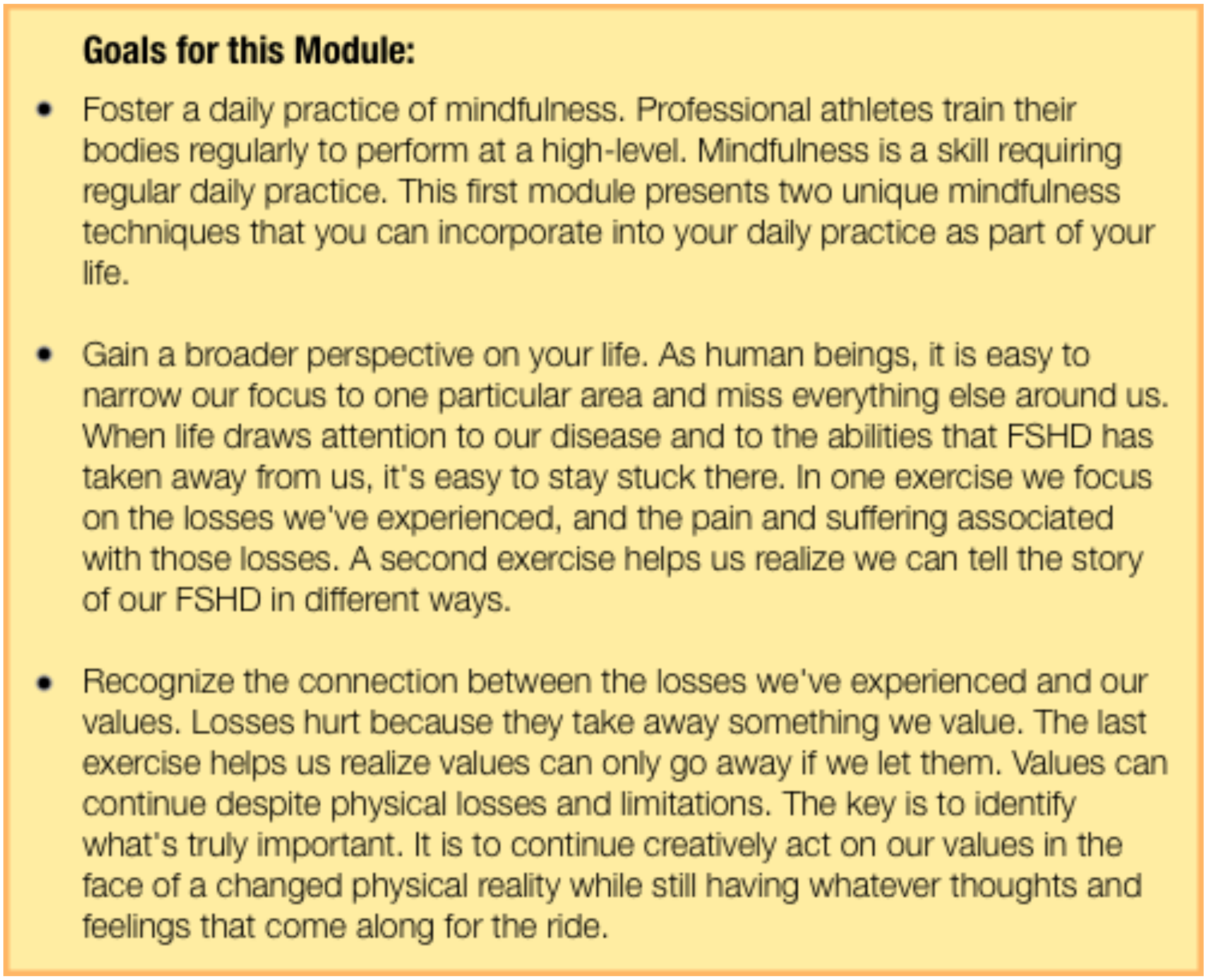
Dealing with physical decline and disability

This module focuses on the mental and emotional reactions that people with FSHD experience when they notice changes in their physical symptoms. Watching your body change and grow weaker can lead to emotional pain like sadness, loss, and anger. On the mental side, changes can occur in how one thinks about one's physical abilities and potential, one's worth or value, and even one's core self-image. Those internal reactions are normal. However, how you choose to relate to those emotional and mental reactions matters. It matters ultimately, because it affects what you choose to do and how you live in the face of these kinds of experiences. What you do affects your ongoing physical health, the quality of your life, and the closeness and intimacy of your relationships. It also may affect your sense of meaning and purpose for living.

Take a few minutes to focus on the experiences described in the two bullet points listed below. Read them carefully. See if you can identify a specific experience or event in your life that corresponds to each statement. Try to keep these personal experiences in mind as you work through this section. As you think about these experiences, note the impact of those experiences in your future life.

- *When something in your immediate experience calls attention to your physical limitations from FSHD.*
- *When something happening now reminds you of abilities or activities FSHD has stolen away. When you powerfully feel the loss of something that mattered.*

Goals for Module 1



Goals for this Module:

- Foster a daily practice of mindfulness. Professional athletes train their bodies regularly to perform at a high-level. Mindfulness is a skill requiring regular daily practice. This first module presents two unique mindfulness techniques that you can incorporate into your daily practice as part of your life.

- Gain a broader perspective on your life. As human beings, it is easy to narrow our focus to one particular area and miss everything else around us. When life draws attention to our disease and to the abilities that FSHD has taken away from us, it's easy to stay stuck there. In one exercise we focus on the losses we've experienced, and the pain and suffering associated with those losses. A second exercise helps us realize we can tell the story of our FSHD in different ways.

- Recognize the connection between the losses we've experienced and our values. Losses hurt because they take away something we value. The last exercise helps us realize values can only go away if we let them. Values can continue despite physical losses and limitations. The key is to identify what's truly important. It is to continue creatively act on our values in the face of a changed physical reality while still having whatever thoughts and feelings that come along for the ride.

Awareness of Current Limitations

As a way of starting, I invite you to close your eyes and simply picture yourself in your mind's eye - see yourself - as in a mirror. *What do you see? Who is that person? What do they look like? How old are they? What are their hopes and dreams?*

I don't know about you, but when I do that, I am completely disconnected from that person I picture from my past. My mental picture of me is stuck in the 1970s. I picture myself in my early 20s, with my whole life still ahead of me. I'm physically healthy and vibrant. I run, play tennis, play golf, and bowl. I've never even heard of FSHD, let alone imagined myself developing it. I have a plan for my life in the future. I know where I'm headed. I'm confident, and my path forward seems easy.

My reality, however, is far different from that. I'm not in my 20s. In fact, I'm a senior citizen. I'm no longer physically lean or strong. My work trajectory changed utterly, and I devoted my life to doing something I never imagined in my 20s. Life has been so much more painful and challenging than I expected. But it's also been much richer. My wife and I have experienced devastating, painful losses of people we cared deeply about. My FSHD emerged, and we wondered what that would mean for us. We've struggled financially, worried about our children, asked whether we made the right decisions and choices along the way. But luckily, those hardships have drawn us closer together. We've experienced the richness of love and togetherness and found friendships that sustained us. We've had amazing opportunities open up for us, experiences that we never could've imagined.

Perhaps you are different. You may have grown up in a family where FSHD was present all around you. Maybe a parent or relative had the disease, and you had a frightening glimpse of what might lie ahead. Maybe you didn't want to acknowledge that you were getting weaker, because you had seen it happen to other family members.

How do you picture yourself in your mind's eye? Our first exercise in this module will be a mindfulness exercise where we focus on the person (and body) that we are and have now.

In Part Two of this book, I noted that living in the present moment is one of the six important core processes of ACT. Mindfulness is a skill we can develop to help us stay in the present moment. Skills need to be practiced regularly in order to be useful. Part Two provided a mindful breathing exercise to introduce the practice of mindfulness. In this module, I expand that exercise to direct your attention toward the parts of your body most affected by FSHD. I changed the exercise below from one published Dr. Walser and Dr. Westrup.[1]

EXERCISE 3.1.1 Mindful Breathing with Imagery

As previously, begin this exercise by sitting upright in your chair, with your feet firmly on the floor, and arms comfortably by your side or in your lap. You may wish to close your eyes to limit distraction. Now, focus your attention on the tip of your nose and breathe slowly. Notice the feeling in your nostrils as air moves in and out. You may sense the air coming in is cooler than *the air as it moves back out from your nose… Gently breathe in and out… Gently observe your breath, paying attention to the sensations as the air flows in and out… If at some point you become distracted by thoughts, take a moment to notice the content of the thoughts, the places where your mind takes you, and then, without evaluating, gently release the thought and return your focus to your breathing. If you get distracted many times, return to noticing your breath each time. Continue to focus your attention on your breath for the next several minutes.*

Now, move the focus of your attention to the rise and fall of your chest as you breathe. Notice how your chest expands and contracts… Notice that you are bringing life-sustaining oxygen into your lungs, which then moves throughout your body… Picture in your mind that you are breathing in this energy as light… Choose a soft color, and imagine that with each breath this colored oxygen fills your lungs… Picture your lungs expanding and contracting and being filled with this soft colored light, and just as oxygen spreads, imagine this light spreading into your heart… bringing warm energy to your heart… And now imagine the light increasing and spreading further throughout your body, just like oxygen would… Picture the light traveling to your shoulder area—an area affected by your FSHD… and growing… so that it spreads down into your arms and hands… Imagine this light flowing upward in your neck and flowing into your head and face, eyes, and mouth… Picture the warm light as it fills your upper body especially those areas weakened over time by FSHD. Imagine the light spreading out to your fingertips and down through your abdomen and hips—continuing down your legs and calves into your feet to the tips of your toes… And now imagine that the warm light can pass outside of your skin. See it spreading until it completely circles you… as if a warm ball of light enveloped you… Allow yourself to just rest in this warmth… breathing in and out… feeding your body with your breath. (Take a few minutes, just relax into this warm colored light)

Now, turning your attention away from the light image, gently focus again just on just the rise and fall of your breath… and then on your body and how it feels to sit in the chair. Notice the placement of your feet and your arms. Picture what the room will look like when you open your eyes and, when you are ready, re-enter the room by opening your eyes.

* * *

This exercise helps you observe your thoughts, emotions, and physical sensations. Practicing it regularly strengthens that ability. Observing these mental processes in the present moment takes you out of autopilot. Awareness in the present moment provides you the opportunity to make intentional choices about your actions. You can move your life in the direction that you choose. It may also help you become more aware of the physical changes in your body related to FSHD. This is important for protecting safety and avoiding accidental falls and injuries. Painful thoughts and emotions may show up when we become aware. Taking actions consistent with our values will often require accepting those unpleasant experiences.

Our Awareness of Physical Decline

FSHD has a way of bringing itself into one's mental awareness regularly. This can happen when one unthinkingly tries to do something (that always used to be easy) and realizes again that it no longer works. For instance, reaching up to get a glass from the cabinet's upper shelf and having it drop and shatter on the floor. Now anyone can drop a glass. When it happens to someone with FSHD, it may reveal unexpected weakness. Immediately, one's attention returns to the present moment to discern if something has changed. The problem is there's no magic gauge - nothing to tell you exactly if or what's changed, and if so - how much. You don't even really know for sure. Other people drop dishes too.

Over time, individuals with FSHD carry an awareness of the various areas of their body that are weak and may interfere with certain activities. People with the disease develop adaptive behaviors to compensate for weak muscles. Often these adaptations emerge automatically, without thinking or planning. However, at some point, the individual's unique adaptive way of performing a task comes into their awareness. The person might see themselves performing that action and notice how different they look from how others look. That awareness often sits in the back of one's mind until something brings it back into focus. Perhaps a stranger asks you why are you standing like that, or walking like that, or comments on how your hands move.

Most of the time, that awareness is not a problem. It's actually a good thing. Having that awareness allows me to think ahead about what I require for upcoming activities. Suppose my wife and I decide to go some place. In that case, it's now automatic for me to think about what the terrain will be like, whether there are stairs or if an elevator is available. How far will I have to walk? Are there places to sit down? Should I wear my leg braces, or should I bring my electric wheelchair? Awareness allows for planning. Planning reduces the risk of falling or of being unable to continue. It's natural that unpleasant thoughts and emotions also show up amid this planning. I now understand the importance of noticing experiences and remaining committed to my values even when feeling uncomfortable.

This awareness of physical change also carries with it some emotional costs. The feeling of being different, of being the object of other people's attention, can trigger emotions. It is also frustrating, discouraging, and exhausting to always have to plan. Having to think so carefully about upcoming activities can reduce the sense of spontaneity. I may avoid certain activities because I don't want to deal with the challenges or be a burden on others. I may do this even when those activities would be meaningful or fun. Ultimately, that avoidance can shrink my life and interfere with my relationships. What might this kind of avoidance cost you? Are you willing to feel these feelings if it meant not having those costs?

The next exercise provides a way to examine the mental and emotional impact that the changes and losses from FSHD have made. I adapted this exercise from one published in a previous book by Steven Hayes and colleagues.[2]

EXERCISE 3.1.2 Your FSHD Suffering Inventory

*I invite you to write a list of all the physical changes related to FSHD that are associated with significant emotional distress. These changes may have come with thoughts that you are broken, or created serious worries about your future. Don't list physical changes that are not associated with emotional and mental reactions. In this exercise, we will focus on how you react mentally and emotionally to the physical changes that come with your disease. Use the left-hand column of the form below to jot down the physical changes. In the middle column, include examples of your reactions. These might include thoughts, feelings, memories, physical sensations, habits, or typical behaviors you engage in (related to the physical change or loss) that may distress you. Finally, note how long you've experienced these physical changes, and these associated mental and emotional reactions in the right-hand column. Don't overthink this task. Just write about the physical changes and losses that bother you most and the emotional responses or the behavioral changes these cause you. Be honest and thorough as you create your "**FSHD Suffering Inventory**" in the space below. After you've completed your list, go back and think about how long these issues have been a problem for you. Write that down in the right-hand column. If you need more space, please use a separate piece of paper to complete this task.*

FSHD losses / changes	Mental/emotional/ behavioral distress reactions I experience	How long this has been the case

Now I invite you to reorganize this list. First, review each of these experiences in terms of their impact on your life and the severity of the distress reactions. Then, in the space below (left column), write the same items, but rank them in order. The order should range from those items that cause you the most pain and difficulty in life to those that cause you the least trouble. You can use this list as a guide throughout the rest of this book. We may ask you to refer to this list as your touchstone for the physical changes/losses that cause an impact on your mental and emotional experience.

FSHD losses / changes	**Mental/emotional/behavioral distress reactions I experience**

Finally, in the area to the right of the list above, draw arrows between every item on the list related to another item. You know that two items relate if changes in one might alter another. For example, suppose one of your reactions to the loss of leg strength is "sadness," and another is "to avoid going out." If you think the two relate (the more sadness you feel, the less likely you will be to go out and be with people, or vice versa), draw a two-headed arrow between sadness and not going out. You may find that many of these things relate, and the space becomes cluttered with arrows. That's fine. There is no right or wrong way to do this. If everything connects, it's important to know that. If some items relate to only a few others, that is useful information too. The higher on your list the experiences are and the more other items they connect to, the more important they become. You may find it useful to consider re-ranking your challenges, and merge some items or, in other cases, divide them into smaller units. If that is so, you can create your final working list below, ranked from highest to lowest in order of impact on *your life. This is your FSHD suffering list from highest to lowest.*

So now you've generated an important list of physical changes related to FSHD that have created significant distress, sometimes for a very long time. Perhaps you've also identified the reactions you've had to those physical changes. You've identified some of the intense emotions you feel, and perhaps even some ways you've tried not to have those emotions. Maybe you've identified meaningful activities that you stopped doing because doing them makes you more aware of those physical changes and losses.

We now turn our attention to our efforts to avoid or control these unpleasant mental experiences that show up. As children growing up in the world, we quickly learn to exert control over physical experiences that cause us discomfort and avoid them. If it's cold outside, sometimes we go into the house to get warm or put on a coat, so the cold doesn't bother us. Many of us also learn as children to control or avoid our emotions. Some of us have been told things like "*stop crying, or I'll give you something to cry about*." Avoiding or controlling our internal experience differs from putting on a jacket to stay warm. Trying to avoid or control our internal experience sometimes leads to unintended consequences.

Dr. Jenna LeJeune and Dr. Jason Luoma have written a recent book about values and psychotherapy.[3] They note that pain is inevitable in the human experience but that people have some choice about the type of pain they will experience. They describe two different pain types. One they call the pain of presence, the other the pain of absence. They describe the pain of presence as "what happens when you choose to live your values." For example, it's the pain that comes when pursuing the value of love in a relationship, when sidetracked by the betrayal of a friend or the death of a partner. The pain of presence becomes possible when we abandon our attempts to control and avoid and persist in pursuing what's most important, even though it makes us vulnerable to loss. In contrast, they describe the pain of absence as pain felt when we are living outside of our values. We experience this pain of absence as emptiness, lack of meaning, lack of direction, and intense regret and remorse for things not attempted.

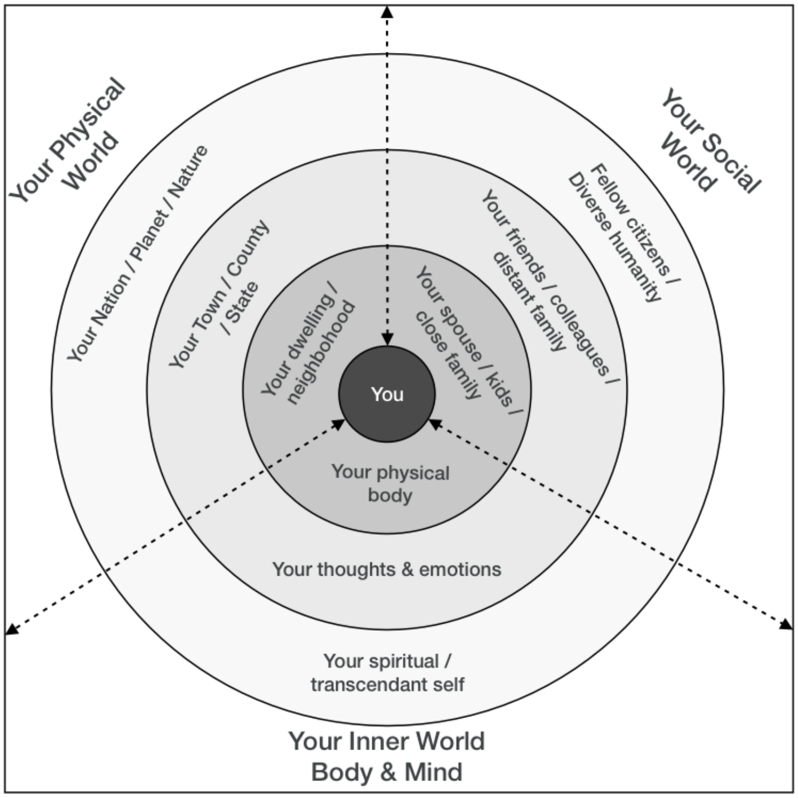
The physical decline, which is a part of FSHD, and losing important activities can be emotionally painful. We may react to that pain by trying to avoid coming into contact with situations that remind us. Yet, in reacting this way, we may end up disconnecting from activities that express important values. We may pull back from life, become isolated, and experience the pain of absence.

This next exercise helps you reflect on important domains of your life and identifies important values. The goal of the exercise is to help you identify actions you can take that express an important value in each life domain. I encourage you to take your time with this exercise. Reflect carefully, and see if you can identify some practical ways to reengage with important values.

* * *

EXERCISE 3.1.3 Connecting to Values

For this exercise, I invite you to look carefully at the graphic below. Notice that at the center is a dark circle with the word "You" inside it. Surrounding that circle are three larger concentric circles. Now, notice the dotted lines with arrows that divide the circles into three segments. Think of these three segments as three major domains in your life. Your inner world is made of your body's sensations, along with your emotions, thoughts, and memories.. Outside of your body, there is your physical world. This is the physical environment all around you, where you have experiences in the present moment. Finally, the third domain is your social world. This includes the people you meet in the physical world, some that you know and love, others that you don't know, and likely some that you don't enjoy being around. As you live your life, you engage and interact, and have experiences within the social and physical worlds. Your mind remembers and evaluates those experiences. It assigns meanings to these within your inner world.



Take a few moments to think about your recent activities in the physical world. Consider things you've done close to where you live or in your neighborhood. Think about something that you've experienced around town or at work, and then even more broadly as you've left your local town to travel to other places. Think about some experiences you've had in the natural world, walking in meadows, forests, beaches, or some other beautiful places.

Now turn your attention to the people in your life and the interactions you've had with them recently. Think about the people close to you who you care about and neighbors and people you've met at the store. Think about people that you've met from far away and notice the strength of connection that you have with each.

Previously, we've described values as important, chosen life directions that guide our activities in the world and our interactions with other people and with ourselves. Bring to mind several of your most important values. Values have no reality simply as ideas. Values only have existence as our actions express them. We can express the same value many times through different actions taken in different contexts.

In the table below, write the values you brought to mind in the left-hand column. In each of the other three columns, identify one or more specific actions you could take to express that value within each specific life domain. I've included a sample in the first line.

Value	Physical world	Social world	Inner world
Health	*Take a walk to a local park. Go for a hike on the trail.*	*Invite a friend to go with you on a walk and spend some time having a great conversation while you walk.*	*Set aside some time daily to establish a mindfulness practice.*

When you've completed the table, pick one or two of the actions, and commit to doing them within the next 24 hours.

* * *

Reminders of Past Losses

The exercise you just completed, hopefully allowed you to think about important areas of your life and ways that you can live your values. We now turn our attention to focus on the role that memory plays in our present moment experience of FSHD. Memory for experiences can be a great treasure but also a source of intense pain. While our actual lives occur in the present moment, we carry our past with us through memory. Most of us would not choose to lose our memory, as it provides a record of our experiences and achievements. We cherish our memories, yet they also bring pain.

Over time, FSHD destroys physical strength and functioning within our muscles. It diminishes or destroys our ability to engage in certain activities. Our memory of lost abilities and activities when triggered can bring pain and suffering back into our awareness. For me, seeing a group of road bicyclists riding together at high speed brings to my mind a sense of disappointment and longing. It brings feelings of sadness, as I can no longer take part in that activity that I loved in the same way. How we engage with the emotions that arise through reminders of losses can affect our experience and our choices. The following mindfulness exercise provides a different way of engaging with emotions that we experience related to loss. Consider your own experiences of loss because of FSHD as you engage in the exercise.

* * *

EXERCISE 3.1.4 Welcome Sadness

Get a pen and paper for this exercise to write your thoughts and reactions. I would like you to start this exercise just as you started the mindful breathing exercise previously. Get seated comfortably, making your posture alert but not rigid.

Gently close your eyes and begin by focusing your attention on your breath. Notice where your attention goes as you do this. For some, you will direct attention to your nose as you observe the air coming into your nostrils, continuing into your lungs, and then being exhaled back out again through your nostrils. For others, you may direct attention to your chest's rise and fall as it expands in and out with the breath. Spend these next few moments being aware of your body and your breathing… Continue following each breath as you slowly and gently breathe in and out.

While you continue to breathe, I invite you to bring to mind a physical change that has occurred in your body related to FSHD. It can be a loss of physical ability associated with the disease wherein you experienced sadness, either now or in the past. As you continue to breathe - focus your attention on that area of your body. Now, bring into your mind an image of that part of your body before the change or loss in functioning. Remember what it was like to experience that part of yourself functioning normally. Now bring to mind an image of how that part of yourself works now. As you breathe, notice what thoughts and feelings are emerging. Notice within you if there is some sadness around that loss of function. Try to allow yourself to hold the sadness gently, as if holding a beautiful leaf in the open palm of your hand.

Now, as you remain aware of your breathing, I would like you to repeat to yourself, silently, on the in breath, "Welcome Sadness," and on the out breath "my old friend." So as you breathe in, say to yourself, "Welcome Sadness," and as you breathe out, "my old friend." Continue to repeat these words as you gently breathe. If you get distracted or caught by a thought or feeling, just come back to "Welcome Sadness, my old friend." Continue to do that as you take 10 to 15 additional breaths.

Now, gently release your attention from the words and your breath, focusing on your body sitting in the chair in this room. Picture the room in your mind's eye. When you are ready, rejoin *the room by opening your eyes.*

Take a few minutes to process that experience. Perhaps jot down a few notes about the thoughts and feelings that came up.

Perhaps sadness is not the primary emotion that you experience when reminded of abilities that FSHD has taken away. You might have different emotions associated with different losses. Some might include sadness, others anger, still others discouragement or hopelessness. If this is the case, I encourage you to redo the exercise using that emotion.

We have very little direct control over our emotions. We cannot turn them on or turn them off at will. They arise spontaneously, or experiences or memories elicit them. They stay for a while and at some point fade away. Struggling with our unpleasant emotions can intensify them and may extend their stay in our awareness. Opening ourselves to experiences can make us aware of having multiple experiences simultaneously. As we loosen the grip on the unpleasant emotion, it frees us to notice other things that are also present in that moment. It also frees us to choose how we will respond to the moment. We can choose to respond with actions that express important values. I would encourage you to make use of this exercise frequently when reminders of past losses bring up difficult emotions.

* * *

Practicing being present and welcoming whatever emotions show up in our lives can pay enormous benefits. Holding those emotions gently, and not grasping them, creates a space for other experiences. This can broaden and even enrich our emotional experience. Research shows that struggling to try to not have painful emotions can backfire, by intensifying them and making them last even longer.

The interplay of thoughts and memory

We experience the world through our five senses. Our brain stores the sensory input from those sensations as memory. In this very moment, as I write, my brain is taking in sensory information. I can tell you what I see as I move my eyes and look around the room. I can tell you what I hear around me and outside, the quiet hum of my laptop, the sound of someone using a leaf blower in the distance, the sudden noise of a car driving by. If I focus, I can notice smells, and perhaps the taste in my mouth leftover from my last meal. I can even feel my skin's sensations as I sit in the chair and place my fingers on the keyboard keys. All of that simultaneous experience enters my brain and is available to be stored. It will stay available to me for a short time in my working memory. If my mind perceives it to be significant somehow, my memory can store it for the long-term. It’s important to remember that memory for an experience in the outer world is not the same as the experience itself. Rather, it’s the meaning my brain attaches to the neural impulses flowing from my senses during the experience. Memories of the same experience may differ from person to person. That’s one reason eyewitness testimony is notoriously unreliable. Different people experience the same event differently.

Let’s assume I can retrieve a memory of my present moment experience of writing. Later, my mind might combine those sensory and other memories into a narrative, a story of what happened while I wrote that day.

Though we may recall specific sensations from particular experiences or events, mostly we retrieve memory as a narrative or a story. Meanings that the mind attaches to the sensations get included as a part of the narrative. If we write the story of a memory, sometimes we can tease apart the facts of the experience and separate them from the assigned meanings. This next exercise will provide you with an opportunity to do just that.

The next exercise is important. It helps us distinguish between our physical experience occurring now in the physical world from our inner mental experience. Our minds evaluate, codify, and attach meaning to those physical experiences. Experiences are just experiences, our minds assign the meaning. This is important because our mental evaluations can sometimes shift our focus and turn us away from the things that matter most in life. Learning to better distinguish our real world experience from our mental world opens up our ability to use ACT processes in ways that may enrich our life.

EXERCISE 3.1.5 Retelling Your FSHD Story

Part One: *Find a quiet, comfortable spot where you can think and write. Take out a blank sheet of paper and a pen (or use your laptop). Write the story of your suffering with FSHD as you might've written it before you began reading this book. Describe the major problems that you've struggled with, and whether they relate to FSHD. As you write about these problems, include the* *historical, situational, and personal reasons that they are in your life and how are they have affected you. Take your time.*

Part Two: *Now go back and read through what you wrote. Underline the objective facts in your story. Facts are descriptions of things, people, or events that have actual existence in the physical world. Facts are not judgments, opinions, or conclusions., do not underline these. Do not underline evaluations about causes. Statements about causation often contain words like "because" or "since.” Now take these facts that you underlined and get a new sheet of paper. Write an entirely different story with a completely different ending using all the same facts. Be creative. Think outside the box. This is not an exercise in evaluating your past or trying to change it. It is just an exercise. See if you can take these same facts and weave them into a very different story.*

Part Three: *Now, compare the two stories. Notice how the meaning of the same facts changes and becomes different. If that doesn't seem clear, take the same facts, and write a third completely different story.*

In having you do this task, we're not trying to show that anything is possible. Nor are we trying to minimize or make fun of the pain and suffering associated with your story. We're really trying to make two points with this exercise. First, is that facts don't determine the story in which we find them. Our minds take facts and weave a narrative and assign meaning to that narrative. Many stories and many meanings are possible using the same facts. The facts of our lives are part of our history and cannot change. However, the story about the facts and our beliefs are permanent only to the degree that we cling to them. Loosening our grip on those stories opens up space for new meanings and perceptions of who we are and who we can become.

Memory and language helped human evolution and advanced technology. Without those two things, humans would live continually reinventing the most essential things. Remembering is essential for daily tasks, like knowing how to do things, where to put items, and basic life skills like cooking and dressing. None of those things would be possible without memory.

My family has experienced the loss of several family members because of dementia. Dementia is terrible because it takes away memory and ultimately makes a person totally dependent on others. Yet, there is a cost to memory. Not only do we remember love and joy and happiness from the past, but we also remember the pain, the losses, and the tragic mistakes of our past. Short of dementia or brain injury, human memory is additive, not subtractive.

We can summon up our memories at will when we choose. For instance, I can decide to think back to when I was seven and remember the experience of joyfully swinging on a swing set or racing my bicycle down the street. But memories also come to us seemingly spontaneously. They can catch us off guard. Often, this happens when one of our senses notice something in our environment that triggers a memory. I smell something, and I am suddenly remembering a fantastic meal with people I love. I see a group of bicyclists riding by, and I'm caught up with memories of my own rides in the past. Memories of precious activities remain when disease takes away physical abilities, but we also know those things can no longer be done. They are gone, possibly forever. Awareness of loss is painful, and simply watching other people can trigger such a memory at any moment. Losses are painful because they connect to things we value.

Two key strategies we learn as children for dealing with our physical environment are to control or avoid it. We learn that if it's cold outside, we can avoid the cold by going inside or controlling our body temperature by putting on a coat. Mostly, avoidance and control work exceedingly well in the physical world. It’s not surprising that we often try to use the same strategies for painful thoughts, emotions, and memories. The problem is that the strategies don’t work very well when applied in our inner world. We often find that they have unintended consequences that make our lives unworkable.

Research suggests that when we try to control or avoid (or not have) thoughts, emotions, sensations, and memories, they actually increase. Trying to not have some inner experience actually makes it show up more frequently. ACT suggests two alternative strategies, instead of avoidance and control. These are willingness and defusion. This exercise helps to connect with values related to physical abilities lost because of FSHD, using willingness and defusion..

EXERCISE 3.1.6 River Rocks

Step One: *For this part of the exercise, I'm going to ask you to go on a brief excursion. Depending on where you live and the landscaping in your yard, this excursion might be to your backyard. But it might also be to a local park or stream. I'm sending you outdoors to find a rock. Not just any old rock, but a special rock. I want you to find a flat rock smooth on both sides, like a river rock. This rock would be perfect for skipping across a lake, though we will not do any* *rock-throwing. Not too big, not too small.*

River Rocks

I'm thinking about a rock about the size of a silver dollar coin. The rock should be large enough to be felt in your pocket, but not so large that you wouldn't want to carry it around. You should be able to hold it easily in your hand and spin it from side to side in your fingers. When you come back inside, wash it off to make sure it's clean and dry.

Step Two: *For this step, I invite you to find a quiet, comfortable place to sit, either indoors or outside. I invite you to reflect on some of the physical losses you have experienced related to FSHD and the impact of those losses. You might take out your FSHD Suffering Inventory and review that. Think about these losses and see if you can identify a single physical loss that has bothered you the most, that is associated with the most distress. Pick a word that captures that loss that's understandable to you. It could simply be the body part where the loss of strength occurred. It could be a word that describes what that body part did previously (the function performed) and can do no longer. It doesn't need to be something that anyone else would understand - just you. Write the word so you don't forget.*

Step Three: *Next, I'd invite you to take some time to reflect on WHY this loss has been so distressing. Clearly, you lost something valuable. The intensity of distress shows the value. What was that thing? If your mind comes back simply to the loss of functioning, I invite you to dig deeper. Why is that loss significant? What activities went away? What did those activities mean to you? How did the loss change your relationships? How did it affect your feelings about yourself? See if you can choose one or two words that capture the important value underneath that change. Write the word(s) down.*

Step Four: *The next step will require that you locate a permanent fine tip marker - such as a Sharpie™. Once you have the marker, take the rock, and on one side of the rock, write the word you picked to signify the loss. Make sure you write large enough so that it's easily readable. Next, turn the rock over, and on this side, write the word(s) that represents the important value affected by the loss.*

*To illustrate, I'll share what I did with this exercise related to a story I recounted earlier. I mentioned that one of the first losses of physical functioning for me was in the extensor muscles of my fingers. When I first did this exercise, the word **"hands"** stood out to me as most associated with the loss. In terms of the value that was associated with the loss. My guitar playing was something significant that I did with my hands that I suddenly no longer could do. I often played guitar to accompany singers at special events such as weddings. Music, for me, was one way of sharing joy with others during important moments.*

*The words that I chose to express the lost value were **"share joy."** The important thing about a value, is that it's never completed. It's not like a task, but you can check off of the list. One can always continue to express an important value.*

Once I dug down and identified the underlying value associated with my powerful sense of sadness, I realized that the value—sharing joy through music is still available to me, even though I no longer play guitar. I have learned that I receive the same personal sense of joy by attending classical guitar concerts and sharing that experience with others by inviting them to come along.

Step Five:

In the last step of this exercise, I ask you to take the rock and put it in your pocket (or purse if you don't have a pocket). Carry it with you throughout the day for at least a week. Keep it near your wallet or keys, so you remember to take it along whenever you leave the house. Throughout the day, stop for a moment and simply notice the rock in your pocket. Picture how it would look sitting in the palm of your hand. Think about the physical loss and the emotions that come up. Hold them gently, as if they were in your hand as well. Then mentally, turn the rock over, see the still important value, and that it still exists. Try to think of an action you could take right now today that will express that value. Then commit to doing it today!

* * *

This module has focused on coming to terms with the reality of physical decline as FSHD progresses. It reflects on the loss of important physical abilities and loved activities that often come with the disease. These losses are emotionally painful because they connect to things that matter deeply to us - things we value. When we experience loss, it challenges us to find new ways to express those values. To do this, we must also carry with us the pain from those losses as we continue to move toward those same values or new ones. We stay stuck in the past with painful thoughts and feelings because we don't want to let go.

The next module focuses on our worries about the disease and what may happen as it progresses. Also, ACT provides a way forward - helping us to get unstuck.

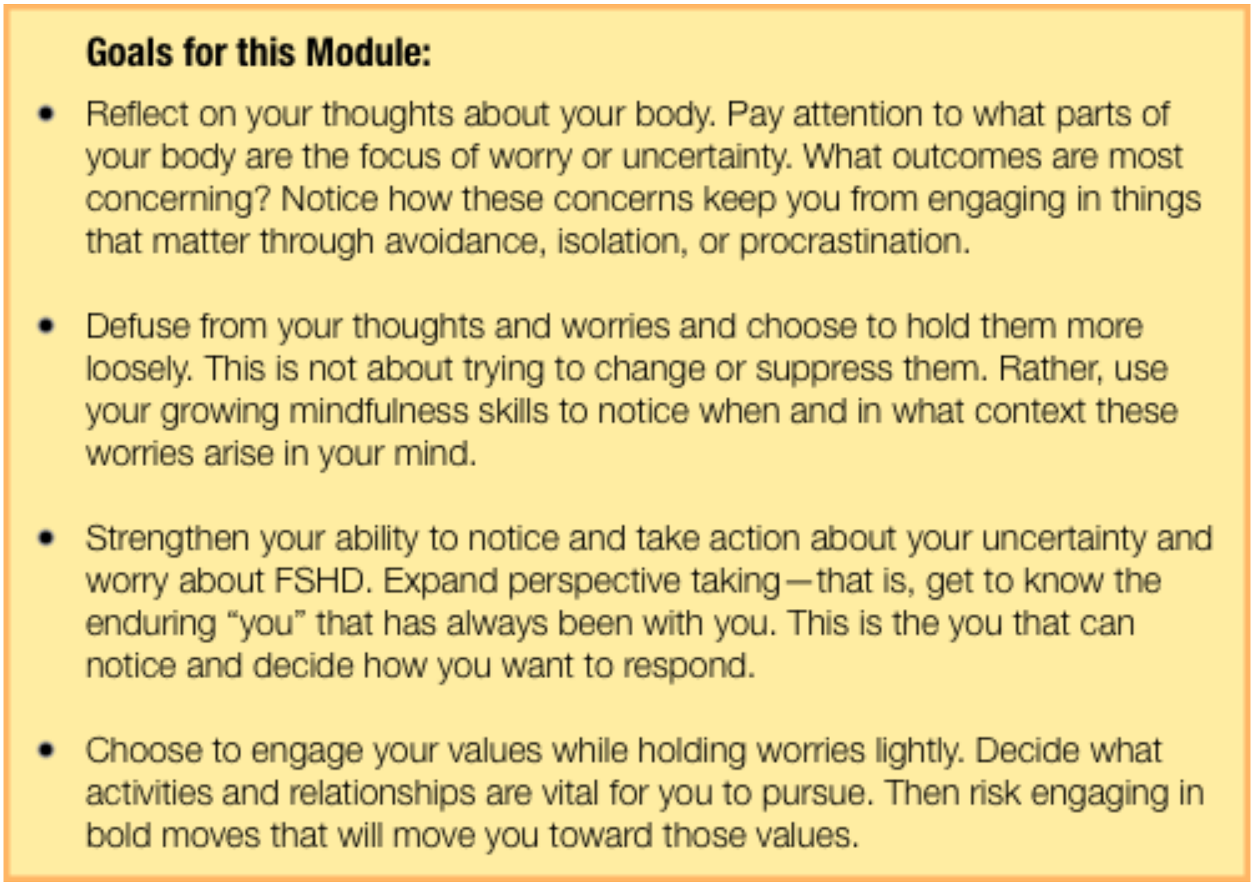
MODULE TWO

Managing uncertainty and worry about FSHD

This module extends our focus on how we engage with our thoughts and feelings about the physical impact of FSHD on our body. We will explore evaluations and the meanings the mind attaches to our disease experience. We will examine two common types of thinking that most of us experience. The first of these are thoughts that arise when we have some unusual physical experience or notice a recent change in our muscle functions. Often we experience uncertainty about what these mean. We have mental questions about the implications of these experiences and how they will affect our life. We also may experience worry about how future FSHD progression might reduce our ability to function. The module looks at thought processes and uses ACT exercises to engage with thoughts and feelings differently..

Goals for Module 2

This module focuses on noticing the thoughts, beliefs, and emotions that show up related to your FSHD. We will practice noticing how these things influence the actions you take day-to-day.



Goals for this Module:

- Reflect on your thoughts about your body. Pay attention to what parts of your body are the focus of worry or uncertainty. What outcomes are most concerning? Notice how these concerns keep you from engaging in things that matter through avoidance, isolation, or procrastination.

- Defuse from your thoughts and worries and choose to hold them more loosely. This is not about trying to change or suppress them. Rather, use your growing mindfulness skills to notice when and in what context these worries arise in your mind.

- Strengthen your ability to notice and take action about your uncertainty and worry about FSHD. Expand perspective taking—that is, get to know the enduring "you" that has always been with you. This is the you that can notice and decide how you want to respond.

- Choose to engage your values while holding worries lightly. Decide what activities and relationships are vital for you to pursue. Then risk engaging in bold moves that will move you toward those values.

Stigma about Disability

As we begin this module, I'd like to return to a word I mentioned in passing in Part Two — **stigma**. The APA dictionary of psychology defines stigma as a *"negative social attitude attached to a characteristic of an individual that may be regarded as a mental, physical, or social deficiency."* We learn stigma from those around us, and once learned, it never really goes away. Much of what we learn in life comes through language - the words we use - and the unspoken messages connected to those words. Political correctness is a hot button topic in our polarized society at present. There has been a good deal of pushback when groups have requested that people stop using certain words or phrases that feel hostile or negative to them. I'm not trying to wade into that debate. Right now I'm not focusing on society. I'm focused on what happens inside my mind - my self-stigma. I'm thinking about the words that come into my mind about me.

I invite you to pause for a moment. Close your eyes and simply think about the words that come into your mind related to your physical disability and appearance. Let your mind run free. Try to come up with every word you've ever heard applied to someone with mobility problems or weakness of some form. If you have a piece of paper, jot them down.

I did this myself while writing. I came up with a few, then looked on the Internet to see what else I could find. I discovered Wikipedia has a whole page devoted to "*disability related terms with negative connotations*." Here's some from my list: Cripple, confined to a wheelchair, gimp, spaz, freak, handicapped, invalid, lame, unfortunate, victim...

Learning is really a one-way street. It's hard, if not impossible, to unlearn things. As a result, the language that we've heard related to disability remains in the back of our minds - hovering. It sits there waiting to emerge. And because the mind links language with experiences that we've had, those words may show up as triggered memories in our present experience. Having learned stigma is a normal experience for everyone. What's really important is how we respond to the negative thoughts and connotations that show up. The first key is to notice they exist, and that they show up from time to time. By noticing, we open up space to choose our response intentionally, and not simply live on autopilot.

Uncertainty in the Present

Not so long ago, I was walking into my house through the back door. There's a single step up required to enter our house. From the driveway, I step up onto a step, and then take another step up through the doorway and into the house. On that day, I tried to step up with my right foot, but it wasn't strong enough. I put my foot back down and tried again using my left leg - and this time, no problem. I was stunned! In 60+ years, I'd never been unable to lift myself up a stair step with one of my legs. In high school, I used to run up flights of stairs two or three steps at a time. So now this happened, but what did it mean?

Forgive me, if it feels like I'm making too big a deal about nothing - but at that moment, it felt like an enormous deal to me. Walking and going upstairs is something I always took entirely for granted. However, when someone has a disease like FSHD, what feels like a sudden emergence of a physical change in functioning can be quite frightening.

Sitting down in the house, I had a flood of thoughts and evaluations. I'm going to take the time to tease apart the various strands of my jumbled thinking - because this is really important.

When someone has FSHD - paying close attention to one's physical functioning as it changes over time becomes important. If I cannot notice what's happening in my body over time, and become careless in my actions, I can end up taking a fall, potentially resulting in serious injury. It is important to be a careful observer of my physical functioning in order to prevent injury. However, our minds also have a tendency to form evaluations about the meaning of experiences. Sometimes our minds attach those perceived meanings to other learned beliefs we carry from growing up, or from other experiences. I know that's vague, so let me just give an example.

So illustrating - let's return to my experience on the doorstep. I've just had the experience of being unable to climb up a simple step. Let's say that my mind leaps to the conclusion that "*I can no longer climb stairs*," and then attaches a label to that experience - "*I've become disabled*." Perhaps most of my previous experiences with disabled people were old people in wheelchairs, shut away in nursing homes - all alone. Perhaps I learned early in life to associate disability with being powerless, worthless, and unable to provide value to others. Trusting evaluations without questioning my mind can drastically affect my life. Holding tightly to those thoughts and beliefs might cause me to disengage from people around me so as not to be a burden. I might isolate myself out of hopelessness and stop pursuing the valued activities that matter most to me.

What I've actually learned is that my physical experience is more nuanced than that. My FSHD has dramatically affected my physical functioning over the last 50 years. The young man that was once vigorous, right-handed and right-footed, is now older, weaker, and now stronger on the left side than on the right because of FSHD. However, I have also learned that changes in my physical functioning aren't entirely linear or entirely one directional. With weight loss and exercise, I gained some strength, but climbing stairs remains difficult and slow. I now realize I can choose my battles with my mobility, sometimes walking, sometimes riding in an electric wheelchair. I am "disabled," but I've also come to realize that the stigma that I learned about disability growing up - doesn't have to be true - even when my mind tells me it is. I am worthwhile, I am valuable, and my presence in the lives of others can bring benefit to all of us.

My purpose in this extensive discussion about thoughts is to distinguish between two distinct thoughts and evaluations. The first type of thoughts - are really observations of the actual present moment physical reality in the world. Noticing that on that one particular day my right leg didn't have the strength to lift my body up a simple step is that kind of thought. These kinds of thoughts/observations are extremely important both to have and to remember related to my FSHD experience. Noticing such a physical experience on a particular day at a particular time can be an important way of marking whether FSHD is progressing or stable. Paying close attention to how my body functions is a way of becoming more knowledgeable about my FSHD experience. Ultimately, this knowledge may protect me from injury.

One function of the mind is to sort quickly through memories, and recall those that seem similar to the present moment experience. It attempts to bring relevant information forward to help us survive and thrive in the world. Mostly, we have little conscious control over what our mind brings forward. It often does matter, however, how tightly I hold on to the information that shows up. It is even more important that I keep the ability to make intentional choices about how I act, and not simply react to my thoughts on autopilot.

For example, when my right foot could not go up the steps - my mind told me I can’t go up the stairs. If I clung to that belief as true without further testing, I might then decide to stop leaving the house to go out. If I were I to do that, my whole life experience and relationships with other people would change, probably for the worse.

In the four exercises that follow, I invite you to continue to explore your inner experience of FSHD. We will begin with a mindfulness exercise, where you focus attention on various parts of your body that FSHD has affected. The next exercise focuses on worry. Worries are thoughts about our possible future that cycle (sometimes repeatedly) through our minds. In this exercise, you can explore a new way to engage with these kinds of thoughts.

In the third exercise, we will ask you to notice the sense of you that continues across time, even as your body changes, and your muscles weaken from the disease. This aware, observing part of yourself is extremely important. This is the part of you that can notice your experience. It is also the part of you with the freedom to choose your actions in the present moment.

The last exercise in this module is an exploration of willingness and committed action. Please use each exercise in the book to improve your skills with the three core practices of ACT.

EXERCISE 3.2.1 FSHD Body Scan

Please begin this exercise by sitting upright and comfortable in your chair with your feet firmly on the floor. Try to be as stable and relaxed as your disease will allow at the moment. Place your arms and hands in a comfortable position, on the chair arms, or in your lap. You might bring a pad of paper and a pen to jot down notes about thoughts or feelings that arise during the exercise. Choose a posture that will allow you to stay focused and alert, yet comfortable.

I would like you to begin the exercise by closing your eyes - or if that's not comfortable - then by finding a place on the floor in front of you on which to focus your eyes. Pause or quit the exercise if any movement causes (or worsens) pain. You are unique, and you are the one who knows your body the best.

During this exercise, I'm going to ask you to attend to parts of your body, one part after another, sometimes asking you to move or tense the area. We will choose areas of the body that are frequently affected by FSHD. As the exercise proceeds, we will focus your attention from one area of the body to another, proceeding slowly downward from your head to your feet.

As we begin, I invite you to turn your attention to your face. Bring your attention to the sensations that you feel on your face. Close your eyes as tightly as you can—hold the tension there for a few seconds, and then relax the muscles. Now, make the biggest smile that you can, until your muscles can't make it any bigger, hold the tension there, and then gently relax your smile. Now do the opposite. Make the biggest frown possible, tense those muscles, then relax again. Finally, tense your jaw tightly, hold the tension as hard as you can for a few seconds, and then release. Now take a moment to imagine in your mind's eye what you looked like, tensing and releasing the tension in those parts of your face. What comes up? What physical sensations did you notice? Were there any particular thoughts that emerged or feelings? Take a minute to jot a brief note about any of your reactions.

Next, I'm going to ask you to focus your attention on your neck and shoulder area. Sit for a minute and scan those parts of your body, looking for areas of tension. Now, tilt your head gently to the left until you can feel the muscles' tension on the opposite side. Now ease your head forward so that your chin comes close to your chest. Notice the sensations. Continue moving your head slowly to the right - tilting it back around so that it is again squarely in line with your shoulders. Feel the tension, now on the left side of your neck as your head tilts toward the right. Now, begin moving your head back so that your forehead faces towards the ceiling. With your head tilted back, continue moving again towards the left, until it completes the circle. Notice the sensations you've been feeling throughout different areas of your neck. Now, return your head to a comfortable, upright, forward-facing position.

Now I'm going to ask you to lift your hands, and slowly raise them toward the ceiling as high as *you can, and hold them there. Notice the sensations. Where do you feel tension? What areas in your arms or shoulders struggle to hold your hands up? As your muscles struggle, you may even notice some gentle trembling. Now, relax your hands and arms and return them back to where they started. Breathe comfortably for a moment. Notice how your shoulders feel now that they're able to relax. Now take a moment to imagine in your mind's eye what you looked like, raising and holding your hands up as high as possible. What came up? Bring your awareness to any thoughts or feelings that emerged as you did this. Jot down some notes about what you noticed.*

Next, I want you to focus your attention on your hips and your abdominal area. First, simply focus your attention and notice the sensations you have in these areas. Take a deep breath into your lungs and hold it for a moment. Notice the expansion of your diaphragm and the tension that builds as you hold your breath. Breathe deeply now for a few moments - paying attention to your abdominal area as you breathe in and out. When you are ready, I'd like you to sit fully upright - as though you were getting ready to stand up from the chair. Now lean your body even further forward—bringing your chest close toward your knees and simply hold yourself there motionless for a moment. Notice the sensations that you feel in your abdomen and back. Now, sit back upright, and then slowly push your back into the back of the chair with as much pressure as possible. Hold the tension there. Notice where you feel the tension in your muscles. Now take a moment to picture yourself standing up straight and tall in your mind's eye. Notice what you look like. What do you observe about your abdominal area? In your mind's eye, slowly walk around. What do you notice about your gait, the way you walk? What thoughts and feeling show up? Bring your attention to whatever emerged as you paid attention to these areas of the body. Make notes about what you noticed.

Finally, I want you to direct your attention toward your legs and feet. To begin, simply notice the sensations. Feel the back of your legs, as they are in contact with the chair. Feel the sensations in the bottoms of your feet as they are in contact with the floor. Now, lift your right foot slightly off the floor and point your toes to the degree you can. Rotate your foot around in a circle. Put that foot back down, and repeat with the left foot. With your feet flat on the floor, try to lift your toes only up off the floor, leaving your heels still in contact with the floor. As you do these actions, notice where in your feet and legs you notice muscle tension. Next, slowly lift your entire leg upward with your knee still bent (as if marching) and hold it raised above the seat for a moment. Notice the sensations and the areas of tension and your muscles, then let your foot slowly return to the floor. Do the same movement with your other leg.

Now tense up the muscles in your quads and calves and hold the tension for a moment—then release it. Notice the difference between the feeling of tension and relaxation. Now mentally picture yourself slowly walking as if in front of a mirror. Notice all the elements of muscle *movement that are engaged in walking. What comes up in your awareness as you think of this? Notice any particular thoughts or feelings that you have as you pay attention to your legs and feet. Write some notes about anything you noticed.*

As we conclude this exercise, I'd like you to just sit for a few minutes and reflect on the scan of your body that you've done. What areas of your body have changed the most from when you first noticed your FSHD until now? Are there movements you can no longer do? Think for a few minutes about the present moment. What parts of your body bring uncertainty at present? By this, I mean, are there particular areas of your body where you worry that weakness is increasing, but you're not entirely sure? Write a few notes about your concerns in these areas. What thoughts and emotions come up when you pay attention to these body parts? Finally, are there other areas of your body where you worry about changes happening that have not yet occurred? Write a few notes about your thoughts, emotions, worries, and concerns for the future.

* * *

What was it like to spend time focused on each of the parts of your body that FSHD affects? I know for me, doing that is not pleasant. I want to turn my attention away. I don't like being reminded of how my disease has changed my body. The exercise we just completed focused on noticing our physical body—especially the parts most affected by FSHD. We also noticed the thoughts and feelings that arise when we pay attention to various parts of our body. It's common for people to go through entire days not noticing very much.

We often live on autopilot—simply going through the motions of living our lives without really noticing what's happening in our inner world. When we do that, we sometimes end up reacting to life experiences in ways that don't work. We may lash out at others in anger when our anger connects to some inner thought we are having, and not what that person has actually done. Misdirected emotional outbursts can harm relationships. Mindfulness is a tool that helps bring us into the present moment. There we can slow down and notice what's present inside of us. We strengthen our ability to notice our inner experience in the present moment each time that we practice mindfulness.

Worry about the Future

The Merriam-Webster online dictionary defines worry as "*mental distress or agitation resulting from concern usually for something impending or anticipated*."[1] In the previous module, we noted how FSHD regularly pulls our attention away from the present moment into the past (through continuing reminders of previous physical losses). However, the disease also focuses our attention on the future (with worries about losses and increasing disability still to come). While this is normal, it is problematic because we can only experience life meaningfully in the present moment.

Each moment we spend ruminating about lost abilities, or that we spend worried about abilities we might lose in the future, is a moment not available for meaningful living right now. The past is gone - we can never truly have it back, and the future does not yet exist. Yet, often we give away our precious present moments. We get stuck in our minds instead of moving about and taking action in the now.

As mentioned, I have a very long history of riding bicycles for fun and exercise. Over the years, I've ridden multi-day bike tours, and century rides (100 miles). I even commuted back-and-forth to work on a bicycle for a time. I've always felt extremely comfortable and confident about riding my bike. A while back, I started riding again after not having ridden for several years. These were short rides on flat roads, less than 5 miles, mostly through residential neighborhoods. On my third or fourth day out, I was nearly home and was crossing a street after stopping at the traffic signal. Halfway across the street, while I was still moving slowly, my foot suddenly slipped off the pedal. I had difficulty getting it back on the pedal while keeping the bike moving. I ended up stopping in the crosswalk, then simply pushing the bike the rest of the way across the street with my feet. It was a little embarrassing, but not really a big deal.

Over the next two days, I noticed myself procrastinating about riding and thinking a lot about the route that I would take. When I finally sat down to analyze what was going on, I realized I was avoiding riding because of fear and worry. On the surface, being afraid made no sense. Nothing bad had happened. But as I took some time to observe my thoughts as they spun around, I realized my fear connected to other experiences the previous summer.

Twice, during trips I had taken that summer, I had stumbled while walking and fallen. However, for the first time, I could not stand back up on my own. I needed help. I suddenly realized that my avoidance and anxiety centered on the fear that I might fall on the bike. I was afraid that if I fell, I might get stuck in the traffic lane, unable to stand up. Now, I've never fallen like that in all the years I have been riding bicycles. But suddenly, worry was overtaking me and keeping me from doing something that I love.

FSHD is a disease characterized by uncertainty. The one certain thing is that the disease will worsen. However, the speed and amount of physical decline are very uncertain. Even among families where several members have the disease, there can be great variability in the age when the disease appears and in its course. Chad LeJeune is a psychologist who has written a book about worry. He defines worry this way: "*Worry can be seen as an internal, verbal process by which we attempt (unrealistically) to reduce uncertainty by exerting imaginary control over the future.*"[2] At its core, worry is an attempt to be in control of a future that feels frightening. But worry is inherently unproductive—it doesn't take us anywhere. It provides the illusion that we're doing something when actually we're stuck. Worry keeps us spinning our wheels and avoiding doing something meaningful. Each moment that we spend in worry is one NOT spent doing what matters and living the life we truly want. So how do we get unstuck? One way is by loosening the grip that our worried thoughts have on us. Doing that takes persistent practice. I adapted the following exercise from one that Dr. Lejeune describes in his book.[2]

* * *

EXERCISE 3.2.2 Keychain Worries

To begin, find a quiet place to sit, take out a paper, and think about your biggest worries related to your FSHD. See if you can identify the concerns you have about the disease that draw you most easily into worry and rumination about what might happen in the future. See if you can identify a single phrase that captures the gist of each worry. Write those phrases down on the piece of paper. Now, one at a time, let each worry play out in your thoughts to its ultimate conclusion: What happens? While you do this, try to notice the thoughts and the emotions that show up. Jot down a few notes about those thoughts and emotions. Spend enough time with this process to identify 3-5 big worries that you have about your disease and its impact on your future life.

Keychain

Most of us carry a key chain when we go outside, either in a purse or pocket. For the next part of this exercise, I'd like you to take out your key chain and look at it. Of course, everyone's key chain is different, but most of us have at least a few keys in common. We usually have a key that opens a lock to the place that we live, and perhaps one or more keys for a vehicle. We might also have an office key, or key to a desk, perhaps key to a padlock - for a bike or a storage unit. If yours is like mine - there may even be a few keys on the chain for which you don't even remember the purpose.

Now, hold your key chain up by the key that you use most often. Hold it in front of your face. Look carefully at the key between your fingers. While you're holding the key, read the first phrase you wrote to describe your biggest worry about FSHD. Read the worry aloud as you look at the key.

Next, take another key that you use frequently. Again, hold it up in front of you. This time, read the next worry statement that you wrote on your paper. While you're looking at this key and reading the statement aloud, picture the outcome that you fear of that worry. Notice that the outcome of that worry hasn't happened in the here and now. Notice your key is real and present in your fingers. The outcome of that worry only exists in your mind. What is accurate right now–in the real world is that you have the power to make choices not about the future, but about the present. You get to decide what you will do today - right now.

When you've finished, pick another key and go through the same process with each of the remaining worries you wrote. You might simply note on your paper which key you associated with each worry. You've now made an association on paper and in your mind between each of those keys and one of your important worries. Notice that you are not the key, and you are not the worry. Instead, you can take the key out of your pocket and hold it in your hand. You can grip it tightly or hold it loosely. You can take it out and look at it, or you can put it away back in *your purse or pocket - it's your choice.*

Notice that there is a YOU that can make that choice. You can choose what will be important to you. And you can choose the actions you will take to live that value—today in the present moment.

I would invite you each time you use each of those keys this week to take a moment to recognize the worry paired with the key. But also take a moment to notice that you are the chooser. In that moment, take some time to choose something you can do today that brings to life one of your most important values. Then do it.

* * *

With FSHD, it's not always easy to know whether fears and worries that emerge are realistic. There's no magic guidebook that can tell you. Changes in physical strength can make previously simple activities hazardous. Maintaining physical activity, particularly when it is meaningful, is important for individuals with FSHD. However, serious falls and injuries can have long-term consequences. The challenge is figuring out the point at which actual risks outweigh the potential benefits. It is important to problem solve whether adaptive changes in how you perform an activity might enable you to continue doing something you love, even if it means doing it differently.

Right now, I haven't made an impulsive final decision about bicycling. I will continue to ride carefully, paying attention to - but not being ruled by my anxiety and worry. However, I will also remain receptive to the idea that I may need to adjust my activities once more to mitigate actual dangers of physical harm. That might include finding a bicycling partner to ride with me. It might mean finding a different bike—maybe one with three wheels, to reduce the risks associated with falling.

The worries you struggle with likely differ from mine. I would invite you to continue mindfully noticing your worries and concerns about your future as your FSHD progresses. Acknowledge the certainty that your body will continue to change while you notice the thoughts and emotions that show up along the way. For things that seem like genuine physical risks, see if you can identify adaptations that will allow you to keep doing the things that are important to you. I would encourage you not to go it alone. Reach out to other people with the disease and to medical professionals for advice. Take advantage of the things their experience has taught them.

Building our ability to be aware of our inner experience, our thoughts, worries, and emotions is a first step in creating a meaningful life with FSHD. We also want to strengthen our ability to shift our perspective - to lift our eyes for a moment and see where we are. The noticing skills you develop through mindfulness are a part of strengthening what we call **our "Observer Self**.”

Have you ever had the experience of walking on a trail with your eyes focused down to keep from stumbling, and then stopped for a moment to look up and outward? Looking up, did you see the stunning panoramic view of nature that you would have missed if you hadn't stopped? The observer self is the part of us that can step back from the immediate chaos of our experience. It is the unchanging part of who we are that is aware of all of our life experiences, including the physical decline of our body with FSHD. This is the sense of self that is constant and continues across time. It is from this place of being able to observe the ongoing flow of experience across time—where we find the space to make intentional choices in the here and now. We can make choices about what is important and choose the actions that flow from those values.

EXERCISE 3.2.3 My Younger Self

This exercise helps you connect with the observer self I just mentioned and the ACT process called "**Self-as-context**" that we introduced in Part Two. It is the aspect of the self that can observe the flow of your moment to moment experience as an ongoing process throughout life. This aspect of self can observe the experience of your bodily sensations. It can observe the ebb and flow of emotions, and it can observe thinking. This is the self that can notice those experiences as they flow ***through you*** rather than ***being you***. This exercise will allow you to practice being the observer.

I often take photos at moments of special importance. Choose two photos of yourself from different times, like one from childhood and one from adulthood. Try to select at least one photo (if possible) from before you noticed your FSHD symptoms. If finding photos isn't convenient, simply sit and reflect on specific moments from those time periods you remember well.

As we begin this experience, settle into a comfortable chair with the photos (or memories). I invite you to breathe mindfully. Focus on your breath, as we have done in previous exercises. Focus your attention on your breath as it flows in and out of your body. If your mind draws your attention away with other thoughts, simply notice that without judging and return your attention to your breathing.

After a moment or two, I invite you to take the oldest of the two photos in your hand and look at it carefully. Notice the you that was then. Notice the clothing you were wearing and how tall you were. Try to remember what was happening at the moment of the photo. What was important to you then? Who were the people in your life? What did you want for your future? Notice how your body differed from your body now. Notice the differences in your level of knowledge and wisdom. Observe the skills that you have now that you didn't have then. Notice also, there may be skills you had then that you no longer have. What thoughts and emotions emerge as you reflect on the you from the past? Jot a few notes down about what comes up. Notice that it is the same you that was there then that is here now in this moment, despite these differences.

Next, take the second photograph, the one taken later in life. Again, try to connect with the person you were in that photograph. What changed as the years passed? How is your appearance different? How did your interests change? What was important to you at the moment that this second photograph was taken? How was that person different from the person who exists now today in this present moment? Importantly, notice that it is the same you that was there then and that is here now in this moment, despite the differences. Again, take a moment to notice any thoughts and emotions that show up about your experience. Take some time to jot them down.

Take a moment to just notice that this observer self is available to you at any time. The observer self can help you step back from your immediate present experience, observe the flow of your experience, and be free to make choices. The ability to choose what's important and what to do next is a person's most important freedom. It allows us to walk forward in life in the direction that we choose. However, to do this requires willingness to carry the other stuff - the thoughts and emotions and sensations that show up and flow through us.

To conclude this exercise, simply return your focus to your breathing. Take some moments just to notice your breath, and when you are ready - open your eyes and return to the room.

Contacting your observer self can assist in your struggle with worry. Uncertainty and worry are often about a conceptualized future self. Worries are guesses about our uncertain future. They often contain our worst imagined judgments of what our life might be like in the future we haven’t yet seen. Occasional worries are normal. Everyone has them. However, when we begin to live and waste our precious present moments there in that imagined terrible future - we cheat ourselves of our precious NOW. We give away our freedom to do what matters right now. Coming into contact with our observer self can teach us that whatever the future might bring. The me that is here now will still be the me that will be there then.

Worry is not the same as planning. Worry is unproductive. It's passive, it doesn't take us anywhere. Planning is active. Because we know we have a degenerative disease, that we will become weaker over time, some planning is appropriate. Planning might involve setting aside money now to pay for expensive modifications or assistive devices. Perhaps planning will mean becoming educated, and educating family members about what might lie ahead and how we might cope. Planning involves taking action now, in the present moment, to prepare for our future, in ways that worry does not. Planning and taking action now can be a way of directing compassion forward toward our future self.

* * *

For many of us, looking at old pictures brings a sense of nostalgia. We remember treasured experiences that were special to us. Some of those memories are happy, some tinged with sadness. I'm able to realize that at least one thing hasn't changed. I am still me! The me that was there before the disease affected my body, that me is still me. My body will continue to change, as will yours. But you will always be there! The amazing part is that we still have this moment right now when we can do the things that mean the most to us. We will continue to explore this topic more fully as we move forward.

Exploring Acceptance

I mentioned earlier people sometimes misunderstand that the acceptance part of ACT when they first hear about it. Some friends with FSHD have initially understood acceptance as giving into the disease or giving up. However, that's not at all what we mean by acceptance. Instead, acceptance is an active stance that I take with my inner experience - my thoughts, emotions, sensations, and memories. Another word for acceptance is willingness.

With either of these terms - acceptance or willingness that stance is all or nothing. There's no in-between. Willingness is not a feeling - it's an action. The act of jumping can illustrate willingness because there's no coming back from it. You can dip your toe into a swimming pool and pull it back, but jumping in means you're willing to get wet. Not only will you get wet, but everything with you will get wet too. In the swimming pool, that might mean your clothes, your wallet, your keys, and your watch. That's because jumping is a committed action in the present moment, and no one truly knows what will happen in the future. We may think we know. Our mind may analyze a situation and predict what will happen. Often, it might even be correct, at other times, not so much. Jumping - like every committed action is an act of faith - done in pursuit of our values. When we jump, we will feel and experience consequences. We may think we know what those consequences will be, but life will sometimes surprise us. Willingness is choosing to move forward with whatever baggage comes along with us. Willingness is taking a step towards something important despite unpleasant feelings or worries.

As my FSHD has progressed, jumping physically is now out of the question. The extra impact on my feet and ankles when I land will almost certainly lead to falling. So as I talk about jumping, for me personally, I'm speaking metaphorically. I'm talking about actions that I can take that feel a little scary or uncertain - but will move me in a direction that matters to me. These are actions that express my values. If physical jumping is out of the question for you, you might use walking as a metaphor for you, or rolling your wheelchair.

Walking is actually quite similar to jumping. Whenever we take a step, all of our weight shifts first to our stationary leg as we lift the other foot up and forward. Then, at some point, as we lean into the step, there's a tipping point that has the same quality of uncertainty as jumping. Each step brings a brief instant where there's no turning back, where all of our weight will inexorably transfer to the foot we've extended forward. We then hope (though without certainly) that our muscles will keep us upright. Walking is an exercise in willingness and committed action, especially for someone with FSHD. Let's take a closer look by completing the next exercise.

Engaging in exercises like the one that follows is like taking that step or making that jump. People with FSHD have uncertainty about their body's functioning and potential changes. It's perfectly natural to have worries about what might lie ahead in the future. Thoughts and emotions rising and falling are a natural part of the human experience. Trying to control inner experience by suppressing thoughts and feelings or avoiding situations where they might arise is often counterproductive. It can even add to our suffering experience. Staying connected to what is most important to us and seeking every day to take part in activities that truly matter is a way to live a purposeful life.

Jumping

EXERCISE 3.2.4 Jumping… Or Stepping

This simple task is an exercise in mindful noticing. There are two versions of this exercise below. Please choose which version of this exercise you will do based on your own understanding of your body's unique strengths and limitations. Please be safe! If you (like me) are unable or uncertain about jumping but can walk (even if it requires a walker/rollator) - please do the Stepping exercise below, where you will take a single step instead of jumping.

Jumping

To begin, find an open flat surface where you are unlikely to fall. Find a safe flat area to stand and breathe as you've done in previous exercises. Now, prepare to jump forward a couple of inches—no need for big exertion. Notice the thoughts that arise in your mind as you ready yourself. Prepare your body to jump forward and to notice all the sensations you are feeling and where those sensations occur. Now jump forward. Notice the tension in your legs as you push upward and forward. Notice the sensation for that moment when your body is defying gravity in the air. Then notice where your body feels the impact with the ground as the burden of your weight transfers upward from the soles of your feet up into your legs.

Now, find a low step-stool or a few books. Place the stool or books on the floor in front of you and step up onto the books or stool. Repeat the process you just did. Notice as you prepare to *jump forward the thoughts that come into your mind. Then jump. Notice how it feels while you're in the air and how it feels on the ground. Notice that in both instances, you made a choice; you acted on it, and you willingly accepted the consequence.*

Stepping

To begin, find an open flat surface where you are unlikely to fall. Stand still (with your hands on your walker if needed) and breathe as you've done in previous exercises. Now, prepare to take a single step forward—no need for big exertion. Notice the thoughts that arise in your mind as you get ready. Prepare your body to take a step forward and notice with your mind all the sensations you are feeling and the places where those sensations occur. Now walk forward one full step. Notice the tension in your legs as you lift your foot upward and forward. Notice the sensation for that moment when your body is defying gravity in the air, before your foot lands and your weight transfers. Then notice where your body feels the impact with the ground, as the burden of your weight transfers upward from the sole of your feet and up into your leg. Notice the sensations as your trailing leg comes forward to join your leading leg.

Please do this next portion only if you can go down several stairs safely without assistance.

Now, find a short staircase with a railing. Stand on the top step, holding the railing. Repeat the process you just did, taking one step forward down the steps. As you prepare to step forward, notice the thoughts that are present in your mind. Notice any thoughts, worries, or feelings. Then carefully step down a step. Notice how it feels while your foot is in the air and how it feels as it meets the lower step. Notice that in both instances, you made a choice; you acted on it, and you willingly accepted the consequence. Hopefully, that consequence did not include falling.

Once you have completed this simple task. Sit down and reflect on your experience. Spend a few minutes thinking about some action you've avoided taking because of your FSHD and the associated anxiety. Maybe it's a situation where you feel a bit out of control or where are you are afraid that others may notice your disability. Choose an activity that aligns with your values but makes you a little uncomfortable. Commit yourself to taking that bold move - hopefully, today or within the next 24 hours.

* * *

As I close this module, I'd like to share with you a metaphor that is used widely in ACT. Its title is "Passengers on the Bus." As far as I can tell, it was first published by Steve Hayes and his colleagues in the first edition of their book.[3]

I rode a school bus for a short time as a child and remember noisy and chaotic rides where the driver had to yell at us to be quiet or sit down. I even remember a day where he stopped the bus and stood up to yell at us. Perhaps you’ve had similar (or maybe different) experiences riding a bus. Try to pull some of those experiences to mind as I share this bus story.

I invite you to picture a bus - with you as the driver. It could be a city bus or something more like a greyhound bus that goes from city to city. Usually on the front of a bus like this, there is a sign showing its destination. On this bus, the sign shows your most important values. The sign shows where you're headed and how you want to be as a person on the way.

Now, picture the bus filled with a bunch of passengers. These passengers are your thoughts, feelings, your bodily sensations, memories. They include your uncertainty about your physical condition and your worries about the future. They are all the aspects of your life experience that live inside you. The bus passengers are your inner world - just as I've written about in other parts of this book.

As you've traveled through life, you've picked up each of these passengers. Some of them you like, and others you're not so comfortable with. Imagine in your mind that some of them look like nice old ladies. They like you and want to sit close to you on the bus. Maybe one represents a thought such as "I'm a good person," or feelings of happiness or contentment you sometimes have. Other passengers look really scary. They're big and look like they might carry weapons. Way in the back of the bus, maybe locked in the bathroom, are some passengers so dark and frightening you don't even know their names. They make you terrified they might come out and do something awful.

What happens is you're driving along and some of the scary passengers threaten you. They tell you what you have to do and where you have to turn. They'll yell really loud - you've got to turn left, or make the next right. If you fail to turn, they get louder and even more threatening. They say things like, "If you don't do what we say, we're going to come up from the back of the bus."

Picture the situation outside the bus. The road is narrow in places, and winding, with lots of twists and turns. There are intersections with cross traffic. If you're not careful, there are genuine risks you could get in an accident, damage the bus and hurt yourself. It's really important that you be able to pay attention to the road and continue driving safely to arrive at your destination.

So you make deals with the threatening passengers, and the deal is, "if you sit in the back of the bus and squish yourself down so I can't see you very often, I'll do what you say. If you want me to turn left, I'll turn left"

Now, what if one day you say “Enough is enough - I'm tired of this! I'm going to throw those people off the bus!” You stop the bus and go to the back to deal with the scary passengers. Except, you notice that the first thing you had to do was stop driving. You notice that now you're not moving forward at all. You totally stopped, and you're just dealing with the passengers. Plus, you discover they're really strong. They don't intend to leave. You wrestle around with them for a while, but they just won't leave. You’re not strong enough to make them go away.

Eventually you go back to placating them again. You try to get them to sit way in the back again where you can't see them. In exchange, you tell them you'll do what they ask just to get them to quiet down. After a while, they don't even have to tell you to turn left any more - you already know, and you do it without them even asking. Eventually, you can get good enough at this that you can almost pretend they're not on the bus. You tell yourself - left is the way you wanted to go, anyway. You're not even going in the same direction as your destination anymore. Instead, you're going round and round in big left-handed circles. Even though the bus is moving, it’s like you’re spinning your wheels. You are not getting any closer to your destination. Maybe you’re even farther away than when you started.

Now, the trick of this whole thing is - the power the passengers have over you - is 100% based on this. **“If you don't do what we say, we're coming up and making you look at us.”** That's it. It's true that when they come up to the front, it appears like they could do a lot more. They show you weapons. They are mean and loud. They look like they could destroy you. But the thing is, they've never actually touched you. They've never physically crossed the little yellow line on the floor that separates the driver's area from the rest of the bus.

The driver (you) has control of the bus, but you trade away your control in the secret deals with the passengers. By trying to get control, you've actually given up control! Even though the passengers claim they could destroy you if you don't turn left, it has never actually happened. The passengers can't actually make you do anything against your will. You still have the choice of where you want to go, and when, and if you turn.

This metaphor is a vivid picture of each of us dealing with FSHD, and everything else that life brings to us. Likely, all of us have spent a great deal of time and effort trying to avoid uncomfortable, even painful emotions, thoughts, and memories. We stop our movement forward and ruminate about the past, or worry about what lies ahead. The progression of FSHD is lifelong, and its course is unpredictable. Each moment we have is precious. If we're going to make progress in moving toward the things that most matter, we need to get back in the driver's seat. We need to restart the bus, and turn the bus in our chosen direction. In order to do that, we have to be willing to have our noisy passengers along with us, even the ones we don't like.

I would encourage you to keep this metaphor in mind as you work through this book and as you live your life. From time to time ask yourself, am I driving toward my destination? And if the answer is no, you need to reset your course, pay attention to the road, and press the accelerator until you move again.

In this module, we have reflected on the relationship we have with our bodies, as FSHD weakens our muscles and makes life activities more difficult. We engaged in exercises to expand our sense of self. We practiced noticing the continuing self that endures across time, place, and the impact of the disease on our bodies. We helped strengthen your observer self, that notices your inner experience, and has the freedom to make choices about how to will live in the present moment.

In the next module, we will examine our moment to moment interaction with our mind and thoughts. We will pay particular attention to the thoughts that arise from our minds when other people see our FSHD - visible in our bodies. We will focus on how we choose to relate to thoughts and emotions about our bodies that are sometimes painful, as well as the choices we make in the face of those thoughts and emotions.

MODULE THREE

Discomfort “being seen.”

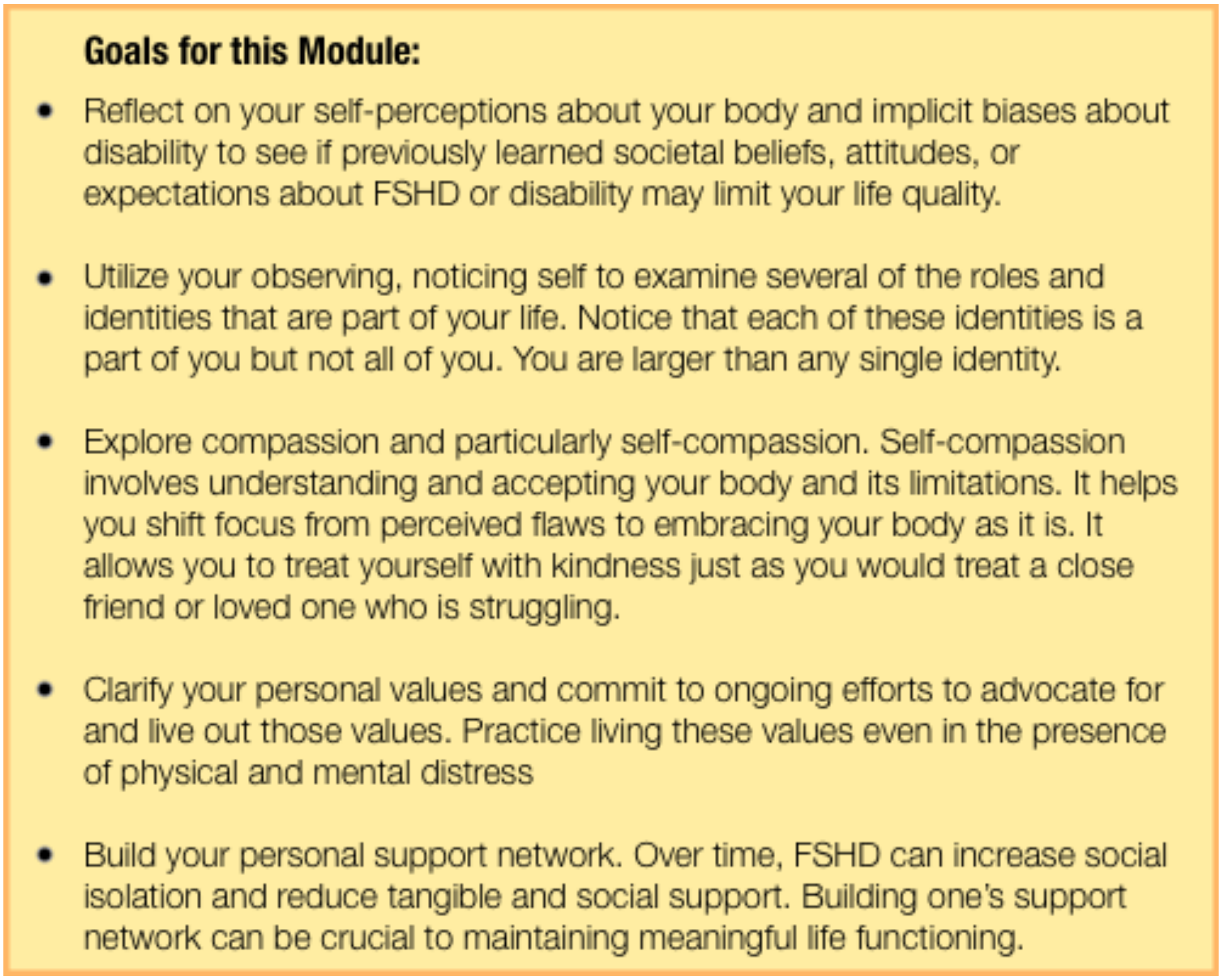
TV commercials and media ads with attractive models show society's high value on appearance. Both men and women in the US struggle with body image because of unrealistic societal standards. FSHD's visible impact on the body can cause individuals with the disease to struggle with their appearance. They may feel distress or discomfort in situations where they feel suddenly "on display."

As the disease worsens and muscles weaken, FSHD becomes increasingly visible throughout the body. Facial weakness can affect an individual’s facial expression, the ability to smile, and to speak understandably. As weakness develops in the shoulders, arms, and hands, normal movements become difficult. Posture, stance, and one’s walking gait may be visibly affected. Individuals often adapt the way they use their hands and arms because of the weakness. Movement in everyday activities may appear unusual or awkward.

Weakened muscles in the abdominal area can cause a protruding abdomen, and spinal curvature called lordosis. These symptoms are quite visible and can also create chronic pain. Finally, foot drop and asymmetrical weakness in the legs and hips can produce highly visible changes in how an individual stands and walks. People with FSHD may feel uncomfortable in public because of their physical appearance.

The PRISM-FSHD study shows how the disease affects self-perception.[1] In this large nationally representative sample of FSHD patients, 92% reported that they experienced body image changes. In terms of the impact of those changes, the study shows that body image changes are the fourth most impactful of all the symptoms listed. Altered body image has a greater impact on patients' daily lives than chronic pain or fatigue.

Goals for Module 3



Goals for this Module:

- Reflect on your self-perceptions about your body and implicit biases about disability to see if previously learned societal beliefs, attitudes, or expectations about FSHD or disability may limit your life quality.
- Utilize your observing, noticing self to examine several of the roles and identities that are part of your life. Notice that each of these identities is a part of you but not all of you. You are larger than any single identity.
- Explore compassion and particularly self-compassion. Self-compassion involves understanding and accepting your body and its limitations. It helps you shift focus from perceived flaws to embracing your body as it is. It allows you to treat yourself with kindness just as you would treat a close friend or loved one who is struggling.
- Clarify your personal values and commit to ongoing efforts to advocate for and live out those values. Practice living these values even in the presence of physical and mental distress
- Build your personal support network. Over time, FSHD can increase social isolation and reduce tangible and social support. Building one's support network can be crucial to maintaining meaningful life functioning.

The following are challenges addressed using ACT exercises during this module.

- Self-consciousness about disability. Being uncomfortable with others watching or noticing physical disabilities. Sometimes this comes about from implicit biases about disability common within our culture.
- Negative thoughts, feeling, and judgements about our appearance can lead us to avoid social situations.
- Over-identifying with one's physical body as the determinant of one's value and/or character
- Avoiding disclosure about FSHD to others. Avoiding potential sources of information about the disease (doctors), leading to lack of access to helpful medical advice or care.

Societal views on disability

Scholars use the term "implicit bias" to understand how discrimination and prejudice persist in society. Implicit bias is learned attitudes and stereotypes that affect decision-making about individuals in certain groups. One way that I have experienced this is in how people interact with me when I'm in my wheelchair, particularly if my wife is with me. Often people will ask my wife if I need something, rather than ask me directly. It almost feels like I'm a non-person, or that they assume I'm unable to speak for myself. Implicit bias operates mostly outside of our conscious awareness. People in a minority group may have biased attitudes and stereotypes about their own group without realizing it.. The research evidence showing implicit bias and its impact on human behavior is quite strong.

Researchers assert that implicit bias is an artifact of human evolution. Humans evolved and thrived even though they're not the strongest, most powerful predators physically. Indeed, part of human thriving came from their ability to form social groups. Humans could find food and protect themselves from predators by cooperating with one another. However, humans originally developed in small community groups, not large, diverse nations. They had to protect themselves from attacks by outside groups of humans and animal predators. Thus, evolution has hard-wired us to be inherently wary of other humans who are unknown or who look or act differently.

It is natural to feel most comfortable around people that we know, or who seem similar to us. We also feel less comfortable around people who are different. Discrimination against differences happens because of stereotypes about race, religion, gender, sexuality, and disability. Being on the receiving end of discrimination, feeling rejected, may cause us to react with feelings of shame and social withdrawal.

What happens then when a physical disability causes a person to be treated as an outsider? Certainly, there is evidence it may affect the person's self-perception. They may experience inner-directed emotions such as shame or outward-directed emotions such as anger. They may experience evaluative, judgmental thoughts about the self. Thoughts such as "*I'm ugly*," "*I'm deformed,*" "*I can't fit in*," or "*It's pointless to try*" may occur.

We all have preconceptions about unfamiliar groups. We may carry negative attitudes and expectations about people perceived to be disabled. For example, in the years since my FSHD became more visible, I've often experienced surprise by how people interact with me. If I am wearing my leg braces on a hot summer day, they are visible because I'm usually wearing shorts. It's startling when people I've never met come up and start conversations with me about my braces. I know that this visible sign of disability raises people's curiosity. Yet, it's quite jarring to me when a stranger asks me intrusive personal questions they would never think to ask some other stranger.

Children are naturally uninhibited. It's quite common for children to point at somebody that looks different and ask their parents in a loud voice, "Mommy, what's wrong with that man's legs?" Mom, of course, will usually look embarrassed and quickly tell the child to be quiet.

I would invite you to think about your own personal experiences of the way people interact with you when they notice your disability. What is your own experience of how people interact with you when your FSHD is visible? What beliefs and attitudes have you learned about disability, and how do they affect your experience of FSHD?

EXERCISE 3.3.1 The *360°* Me

To complete this exercise you will need a camera or smartphone that can record video. You will also need a tripod. If you don't have a tripod, you will need to recruit a helper to assist you and with the recording. Next, you will need to find a flat level place free of obstacles where are you can walk safely (or roll) 10 or 20 feet in one direction. Please use whatever mobility devices you would normally use. If you use a cane, or a walker - use them. If you no longer walk safely, do this same exercise from your wheelchair. The exercise will involve moving back-and-forth in front of the camera.

When you are ready to begin, set up the camera / tripod at one end of the walking area. If you have no tripod, have your assistant prepare to record you. Set the zoom on the camera so that, for most of the distance, the camera will record your whole body as you move.

Start the recording. Now, walk (roll) at a normal slow pace away from the camera for 10 to 20 feet. This exercise has two parts and will involve both willingness and noticing.

As you get ready to begin, take a deep breath. Then pay attention to your thoughts and feelings, and continue to do so throughout this entire process. Start the video recording. Begin walking (or rolling) slowly away from the camera. When you've gone 10 to 20 feet, stop. Turn around in a circle, a full 360° so that the camera sees every part of your body. Now, turn back around and *face the camera. Then, walk slowly (or roll) back toward the camera until you get to the place where you started. Make sure not to rush. Don't fall. Take your time. Pay careful attention to any thoughts, emotions, evaluations, or physical sensations that emerge so that you can jot them down later. Stop the recording.*

Now, take the smartphone (and your helper if you have one) and return to some place where you can sit comfortably and reflect. Play the video. Carefully observe your body in the video as you slowly walk away from the camera and then turn 360° around. What do you notice about your stance, your posture? Which parts of your body are your eyes most drawn to? What thoughts emerge as you watch the video? Do you experience any emotions while watching? Do you notice any self-stigmatizing thoughts or beliefs about your body?

If you have a helper, or if you're able to get somebody to watch the video with you, ask them what they notice - what stands out to them. Ask them what thoughts and feelings they experience watching the video.

Now watch the entire video again. In the video segment where you are walking away from, and then back toward, the camera. What do you notice about your gait walking away from the camera, and then turning and walking toward the camera? If someone is with you, ask them what they notice. Note any thoughts, emotions, memories, or sensations you each experience while watching the video. Take a few minutes to reflect on your personal observations. What are the stigmatizing thoughts or biases that arise in your mind that you learned from society?

Jot down some notes about what you experienced. If you're willing, talk with your helper about what the experience was like for you, and ask them what they noticed. How were your observations similar or different?

Many people (not just those with FSHD) feel some discomfort seeing themselves in video or photographs. Part of this is that it's not the experience that we usually have. We can't physically see ourselves from a distance, we usually only see ourselves in mirrors, where our image is reversed. Your helper likely observed the you they always see. How are the things that you noted similar or different from the things that your helper noticed? Talk together for a few minutes about your experiences.

Now, imagine that the video you just watched was of another person, perhaps someone you know - a friend or a loved one. Notice any compassion you might experience in that situation. See if it might be possible to direct that same compassion toward yourself.

If stigma or bias showed up for you during the exercise, notice that you learned these biases. While we can't unlearn our biases, we can decide to loosen the grip they have over our actions. Notice that you can have these biased thoughts and still choose to engage in actions that express your most important values.

* * *

Identity and Self-Image

In college, I often avoided studying by read*ing* J.R.R. Tolkien's books. One of my favorite passages from the Hobbit occurs when Bilbo hides, trapped in the treasure chamber of the dragon Smaug. The dragon knows that he's there but can't find him. The dragon calls out to him and asks about his identity. Bilbo answers with a riddle. He says:

> *"I come from under the hill, and under the hills and over the hills my paths led. And through the air, I am he that walks unseen. I am the clue-finder, the web-cutter, the stinging fly. I was chosen for the lucky number. I am he that buries his friends alive and drowns them and draws them alive again from the water. I came from the end of a bag, but no bag went over me. I am the friend of bears and the guest of eagles. I am Ringwinner and Luckwearer; and I am Barrel-rider."*[2]

If you are familiar with the story, each one of those statements connects to something that happened to Bilbo in the story. Each is a clue to his identity. As we live our lives and have experiences that are both joyful and painful, our minds also shape a narrative of our identity. For those of us with FSHD, the way we perceive ourselves with the disease likely affects our personal narrative.

As is clear in the Tolkien quote, people often identify themselves by what they do, where they come from, by other people they know, or by some special quality of their character. We all have many identities or selves. One thing I learned in childhood was about shaking hands and introducing oneself to someone else. Having a firm handshake is important for men, but my FSHD symptoms made it challenging and sometimes awkward. As I grew up, I also came to realize that one of the first things people ask when they meet someone new is "*What do you do?*" In our society, traditionally men define themselves and their worth by what they do. Worth is often associated with physical strength and with the status of their employment.

I've mentioned that one purpose of ACT is to help increase our psychological flexibility. One way of doing that is to come into contact with a sense of self that is larger and separate from any single identity. When we step back and connect with a bigger sense of self, thoughts and feelings become less threatening since they don't define us. We have thoughts and feelings, but we are not our thoughts and feelings - we are much more. Being aware of that larger sense of self - frees us to choose who we want to be, and how we want to act in any situation. I have changed the next exercise from one first published in Dr. Walser and Dr. Westrup's book.[3] It focuses on being able to step back into our observer self to examine and hold lightly our various roles and identities.

EXERCISE 3.3.2 Holding Identity Lightly

Instructions: This exercise strengthens your connection with your observer self. Make sure you have available about thirty to forty minutes to complete the activity. Find a place quiet and free from disturbance. Choose a place where you can reflect. Read the following and then complete the writing and imagery exercises. Begin this exercise by breathing slowly and steadily, focusing your attention on your breath. As you breathe, notice the flow of your thoughts and feelings throughout this exercise.

Sometimes, when we get hooked by our thoughts or emotions, we fuse our focus to a particular aspect of ourselves. It's as if we have welded ourselves to one specific identity, as if there were no other way to view ourselves. For example, suppose you experienced a powerful trauma in your past. In that case, the identity of being a victim might become so predominant that other aspects of yourself get lost. We can do this with many kinds of self-perceptions or identities. You can have a self-concept as a professional, as a strong or a weak person, or as a victim, a survivor, a dad or mom. The list can be long. When we can step back and take a broader perspective as the observer self, we can understand that every self-concept or identity is to be held lightly. None of these are fully complete or perceived as fully, literally true. The aim of this exercise is to contact the sense of self that is larger than any single identity.

I'd like you to begin by writing a description of yourself as each of the identities listed below. Include what that identity would think, feel, sense, and look like:

Your work or school self

Your FSHD affected self

Your best self

Your playful self

Your struggling self

Your ________________ self (one of your choosing)

After you have a brief description of each of those identities, find a quiet place to do a short imagery exercise. As we did previously, take two to three minutes to notice your breath, allowing yourself to observe your breath as it flows in and out of your body. Then, quietly shift your attention to your imagination and picture each of the identities that you have written about, one at a time.

As you picture each identity individually, imagine what that identity might say to you. Ask yourself the degree to which stigma in bias you have experienced he plays a part in that identity. Ask whether stigmatizing beliefs and attitudes you have learned growing up are a part of that identity. Have your image of that identity talk to you as you practice just observing what is being said.

*Now, picture yourself holding a small version of that identity in your hand. As you do this, say to yourself, "**I hold this image lightly, like a warm breeze carries a small floating feather. This identity is not fully me anyway**" Do this with each of the identities. After you have gone through each identity, say to yourself, "**I hold ALL my identities lightly, like a warm breeze holds a small floating feather. They are not fully me, anyway**."*

Then gently focus your attention back to your breath, taking a couple of minutes to notice your breathing. Then turn your attention back to the room where you have done the exercise by gently opening your eyes.

Take a final look at what you have written related to each of these identities. Each of these identities has been shaped by past experience, some or all may be influenced by learned stigma. Note that each of these identities is a part of you but not all of you, and that important values are connected to aspects of each identity. Finally, Focus on the part of you that exists separate from any single identity - the part of you that notices and can decide in any single moment to actively express those values

Take a few moments to jot down some notes about your experiences and reactions to the exercise.

* * *

Understanding our identities and how we move between different roles and activities is important. Our ability to step back into the observer self can aid in awareness of how our self-perception and roles connect. When in contact with that larger sense of self, we can make choices about what matters to us right now, and choose values-based actions in the present moment.

I encourage you to remember this exercise. Try to notice situations when one or more of those roles or identities are present in your life in the coming weeks. When awareness of your FSHD brings up uncomfortable feelings, try picturing a small image of your FSHD self in your outstretched hand and hold it lightly.

That we have unique identities that emerge in situations where we carry different roles and responsibilities is not a problem. My role as a psychologist requires me to prioritize client needs over my personal life. I attempt to keep my other selves outside of the therapy room. This is a good thing for my clients, as it helps keep therapy focused on their specific issues and needs instead of mine.

As my FSHD symptoms became more visible, occasionally a client would ask me a question about a particular symptom that they noticed. This would create a clinical dilemma for me. I would need to decide whether, and how much, to disclose about my FSHD to my client. Unless the client was using the question to avoid dealing with their own struggles, I would often briefly tell them about my disease. Sometimes, I found the clients would start seeing me as a person who struggles like they do, and find my disclosure deepened the clinical connection.

In many workplaces, outside identities must be kept separate by being careful about sharing personal information..

That being said, there can also be painful consequences when we attempt to hide aspects of our self. When we avoid disclosing our internal struggles with FSHD from others, it prevents them from understanding us more deeply. This is especially true when we hide from the people who care most about us or from those who may have access to helpful resources. When our internal struggle causes us to avoid disclosing our disease struggles and our fears to others, we cut ourselves off from support and help. When our fear of being seen keeps us from asking for help when we need it, we limit our options and may even increase our risk of injury. Avoiding contact with painful thoughts and feelings that we carry, and hiding our true selves, has consequences. It can cut us off from people and resources that could make life more meaningful.

Avoiding painful feelings and hopeless thoughts about how FSHD has ravaged our body and changed our life can lead us in unhelpful directions. It may have the unintended consequence of preventing us from accessing the very things and people that might improve our experience.

Have you ever seen a child, hiding and afraid and felt compassion for them? Seeing such a child often provokes the desire to take them in our arms, hold them tightly, and tell them, "*It's OK—it'll be all right!*" There's a sense that each of us with FSHD is like that child. We hide ourself - afraid to let others know how much we are hurting. This next exercise provides an opportunity to contact that hiding inner self and respond to that self with compassion.

EXERCISE 3.3.3 Compassion Mindfulness

We will start this exercise by getting comfortably settled in your chair. Close your eyes and begin by focusing your attention on your breathing. Notice the cool air as it enters your nostrils and fills your lungs, and then notice the warmer air as it leaves your body. Spend a few minutes being aware of your breathing ... Experience each breath as it flows in and out. (Allow a minute or two to focus on your breathing.)

Now, I would like you to shift your attention and spend a few moments thinking back on some particular aspects of the struggle you've had concerning your FSHD. It might come into your awareness as some beloved activity that you no longer can do in the same way. You might notice your frustration at the difficulties involved in getting to places you would like to visit. It might only be a sense of how hard life has become. Pause, and just notice how long you've been experiencing this struggle with your body.

Notice the painful emotions and the wishes that you have that things might be different. Notice the harsh judgments you may have directed toward yourself and your body and how the disease has affected the way you see yourself. Pay attention to beliefs and attitudes you've learned about disease and disability. Notice how those thoughts and beliefs might connect to the way you see and feel about yourself. As you continue to look back, think about how much you have needed others to understand your struggle and accept and love you for who you are. Try in your mind to imagine what it would be like to feel fully accepted, understood, and loved.

Now, imagine that you are as large as the universe and that you have all the capacity to hold these struggles cupped in the palms of your hands. Envision that you can give all the warmth, acceptance, and love that you needed then and that you still need now. Picture yourself as big as the universe and embrace your body struggle with a warm blanket of acceptance. Imagine that you could let your body be just as it is today, yet tightly wrapped with a blanket of acceptance.

Gently hold it in your outstretched palms with compassion. Imagine yourself holding your body and your struggles gently with compassion. Imagine that you can hold and still have this *struggle and still be 100% acceptable and lovable. For these next few moments, see if you can experience yourself as whole. Spend a few moments noticing what that experience of wholeness and completeness is like. Notice that you can offer this compassion to yourself amid any experience.*

Now, gently focus your attention again on your body sitting in the chair, sitting in this room. Picture the room in your mind's eye, and when you are ready, rejoin the room by opening your eyes.

◆

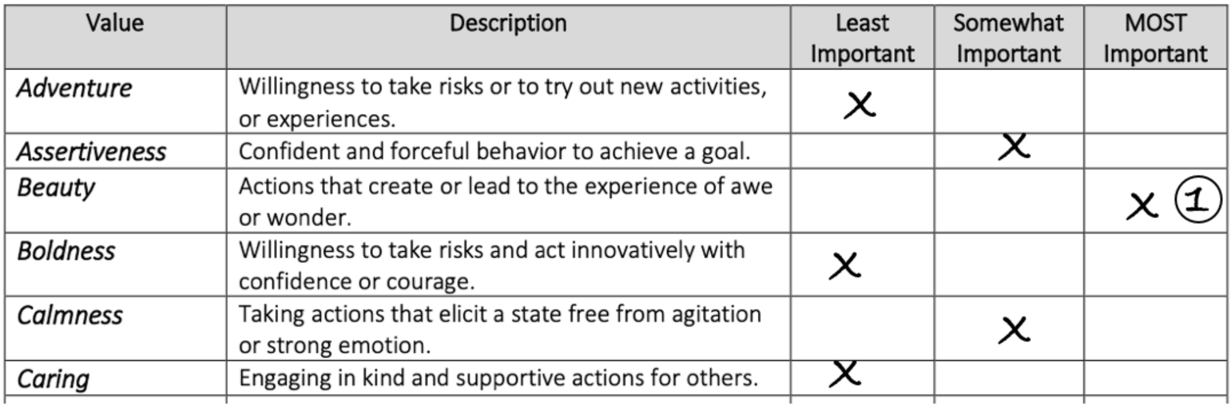
There is nothing better than snuggling into a warm blanket on a cold evening. Self compassion is like that. It can provide warmth and comfort within ourselves to keep moving forward with our disease. This can be true even in situations where we feel uncomfortable being noticed by people around us because of stigma and bias that we have learned. Moving toward the things that matter brings vitality and meaning to life. Identifying the things that matter most to us this is the first step in taking actions that bring our values to life.

This last exercise provides an opportunity to reflect on what values are most personally important for us as a person with FSHD. Knowing what's important and prioritizing our values is a way of moving forward on our chosen path. This is part of moving in your chosen direction. I hope you find this next exercise useful in identifying your most important direction and prioritizing your next steps.

EXERCISE 3.3.4 Values Sort

In **Appendix 5**, at the back of this book, you will find a longer form that looks like the example below. This example shows how to fill out the form found in the appendix. This exercise has three steps.

EXAMPLE:



Value	Description	Least Important	Somewhat Important	MOST Important
Adventure	Willingness to take risks or to try out new activities, or experiences.	X		
Assertiveness	Confident and forceful behavior to achieve a goal.		X	
Beauty	Actions that create or lead to the experience of awe or wonder.			X ①
Boldness	Willingness to take risks and act innovatively with confidence or courage.	X		
Calmness	Taking actions that elicit a state free from agitation or strong emotion.		X	
Caring	Engaging in kind and supportive actions for others.	X		

Step One: *Now, using the full form from* ***Appendix 5****, carefully read through each of the 44 values listed. Notice that some values may feel more important in situations where bias, stigma, or discrimination exist. When you have read them through - return to the beginning. Carefully examine each value on the form one at a time and decide which of the three columns on the right that value best fits for your life with FSHD. Pay particular attention to the importance of each value may be in situations where you are aware of stigma related to your disease and even discrimination. If the value best fits "****Least important****" place an* ***X*** *in that box. If the value is one of your* ***Most important*** *values, place an* ***X*** *in that box. If the value has some importance but is not one of your most important value then place an* ***X*** *in the middle column.*

Step Two: *Once you have placed an* ***X*** *in one of the three columns for each value on the list, carefully examine each of the values that you have selected as your most important values. Please identify your top 5 values and rank them from highest to lowest by placing a circle with a number next to the* ***X*** *in line 3. If the most important list contains over five values, just rank the top five. If you have fewer than five values listed as most important, simply rank each of them from highest to lowest.*

Step Three: *Reflect on each value that you ranked as* ***most important.*** *Try to identify one or two specific actions you could take today that would express that value. Try to consider actions that might be especially relevant in situations where disability bias is present. Think particularly about actions you might otherwise avoid - because it calls attention to your FSHD, and elicits uncomfortable feelings. For example, in the sample above, "****Beauty****" is the most important value. There could be several actions you could take that would bring you into contact with or express beauty. However, some of them might require using assistive devices, or asking for help in ways that might feel uncomfortable. You could go with a friend to an art museum and appreciate the artist's skills and the meaningfulness of the content. You could travel to a beautiful location around dusk and sit and simply watch the sun set in the sky. How you choose to express the value that is most important to you is up to you. Work through each of the 5 most important values that you ranked and identify an action that you can take to express each* *of those values during the next seven days.*

* * *

This module focused on the thoughts, emotions, and memories associated with our physical appearance and 'being seen' by others. It focused on noticing the thoughts and emotions that show up in those situations. We strengthened our ability to hold our multiple identities lightly, recognizing that none of our identities encompass all of who we are. We engaged in a mindfulness exercise where we directed self compassion toward ourselves and our bodies. Finally, we examined the values that we choose to prioritize and worked on identifying tangible actions we can take to live those values.

As we conclude this module, is important to recognize the powerful impact that bias and stigma have on the way that we see ourselves. Bias can come from within or from others' discrimination and stigma. Differing from others because of FSHD can leave us feeling isolated. Feeling rejected by self and others can lead to withdrawing from previously enjoyable activities and people. Yet, social support and connection are the source of tangible and intangible resources for a living meaningful life.

In Part Two of this book, I described an exercise called *Bold Move* (Exercise 2.7). I would encourage you now to return to that exercise and see if you could identify a values-based bold move you could engage in today that would enlarge your support network and strengthen your relationships with family and community.

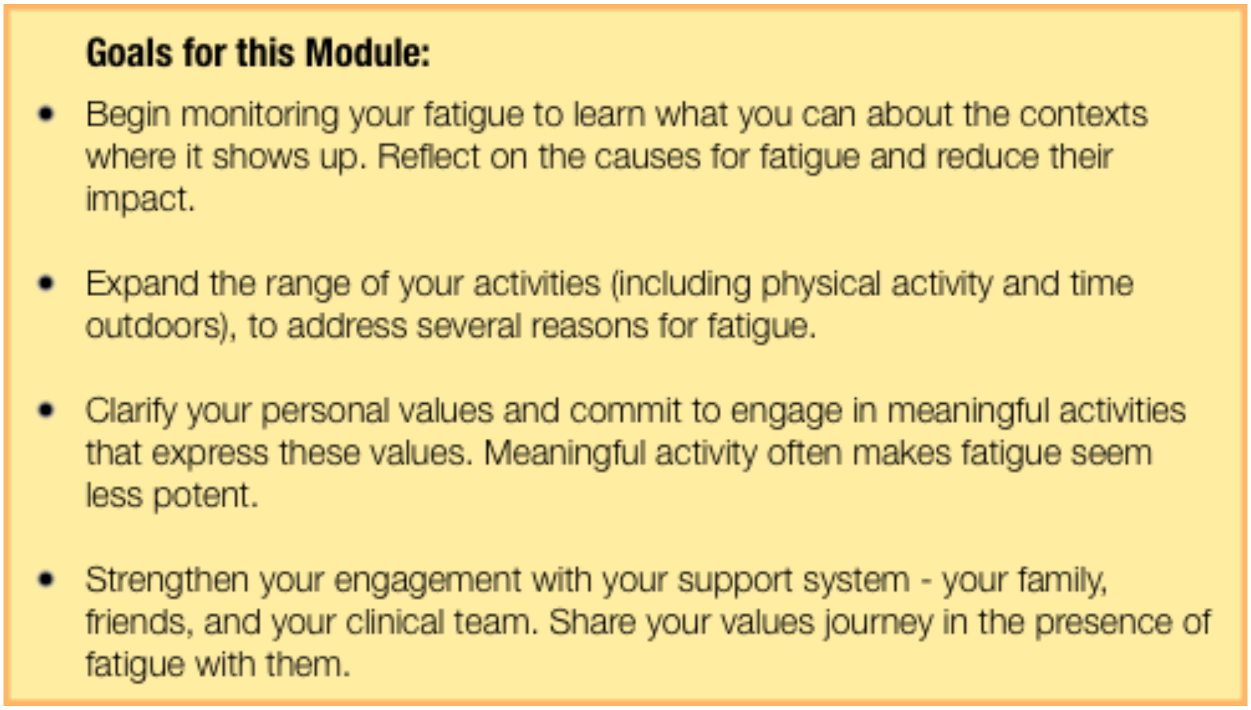
In the next module, we will take up the challenge of fatigue with FSHD. We will return to some of the research findings related to physical activity and fatigue. And we will also explore potential individual sources of fatigue. We'll use mindfulness to observe our thoughts and emotions when fatigued. We'll also explore the importance of social connection and physical activity.

MODULE FOUR

Managing chronic FSHD related fatigue

This module focuses on the fatigue that is reported by a high percentage of FSHD patients. Fatigue can arise from many sources. However, when fatigue becomes chronic, it can become highly debilitating. It can seriously impair your quality of life and affect your normal functioning at work, school, or other important activities.

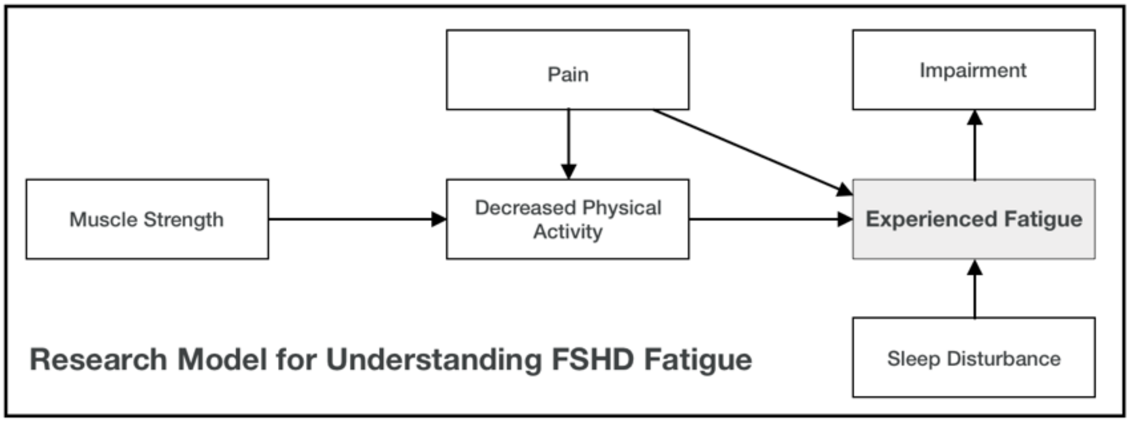
Goals for Module 4



Goals for this Module:

- Begin monitoring your fatigue to learn what you can about the contexts where it shows up. Reflect on the causes for fatigue and reduce their impact.
- Expand the range of your activities (including physical activity and time outdoors), to address several reasons for fatigue.
- Clarify your personal values and commit to engage in meaningful activities that express these values. Meaningful activity often makes fatigue seem less potent.
- Strengthen your engagement with your support system - your family, friends, and your clinical team. Share your values journey in the presence of fatigue with them.

While there is a good deal of research on chronic fatigue unrelated to FSHD, there are only a few studies on fatigue associated with FSHD. As mentioned briefly in Part One, Dr. Kochman and colleagues created a research model to study fatigue in neuromuscular diseases. They also developed a specific model for FSHD.[1] The graphic below shows that model.



Examine the graphic. You see three arrows leading to the slightly gray box that says experienced fatigue. These are the three things most associated with fatigue in FSHD. These are pain, decreased physical activity, and sleep problems. Pain also leads to decreased physical activity, as does reduced muscle strength. FSHD causes muscle weakness, which often results in **less** physical activity. Though it seems counterintuitive, it is actually **too little physical activity** (as opposed to too much) that is one of the direct causes of fatigue.

Second, fatigue can also result from sleep problems. Chronic sleep impairment will often lead to daytime fatigue. Getting enough sleep is a massive problem for much of the US population. There is extensive literature about ways to improve sleep. One way to better understand the impact of sleep on fatigue is to self-monitor. In **Appendix 8** you will find an FSHD Sleep Diary form.

Muscle weakness from FSHD can make it challenging to move about and roll over in bed. Staying in one position for an extended time can lead to numbness sufficient to wake someone up from sleep. Pain-related to the disease can also interfere significantly with sleep.

Sleep issues can also stem from changes and problems with our internal (circadian) clock. Travel (jet-lag), work schedules, too much screen time, and sometimes just the change in seasons can affect sleep. Spending more time outdoors in the sunshine can help reset our internal clock and ultimately reduce fatigue.

I would strongly encourage you to print a couple copies of **Appendix 8** to keep beside your bed. Take a few minutes and complete this form each morning for a week or two. Pay particular attention to the Comments box. Use this box to note anything that occurred during the night that contributed to the Quality of your sleep. This could be noise or distractions that occurred. It might include multiple trips to the bathroom, or physical pain during the night. Particularly jot down any FSHD symptoms that contributed to impaired sleep. Doing this will give you a more complete picture of the role sleep problems may play in your fatigue.

Sleep apnea is widely prevalent in the US within the general population, and some individuals with FSHD will have it as well. People with FSHD may also have additional breathing-related problems that affect sleep. These stem from weakening muscles in the abdomen and diaphragm. If it appears sleep is contributing to your fatigue, consider requesting an appointment with a sleep specialist. for assessment and possible intervention. The doctor may suggest a medical or behavioral intervention after checking for a sleep disorder.

Finally, if you examine the graphic above, you see that pain plays a role in two ways. Physical pain contributes to decreased physical activity, and pain is also a direct cause of fatigue. I will address FSHD-related pain more fully in the next module.

As mentioned in Part One, Dr. Voet and colleagues studied two interventions designed to decrease FSHD patients' fatigue.[2] Both interventions attempted to help FSHD patients to increase their overall physical activity. One intervention comprised 16 weeks of aerobic exercise (40 minutes three times per week, one of those sessions with a physical therapist). The second intervention comprised between 3-6 fifty-minute sessions of individual psychotherapy (cognitive-behavioral therapy-CBT). The goal of these sessions was to reduce fatigue and increase physical activity by addressing negative thoughts and lack of support. Both these interventions were equally effective in reducing fatigue. However, the CBT intervention reduced more of the factors known to be associated with fatigue than did the exercise program.

Though not included in Dr. Kaulkman’s model, research in the general population shows that fatigue can result from mental causes. For example, both sleep problems and fatigue are diagnostic symptoms of depression. If you're feeling down or have lost interest in things you used to enjoy, it's important to evaluate depression. There is a simple screening test for depression included in **Appendix 2** of this book that you can take. If your score falls in the depressed range, it would be worth consulting with a mental health provider or your doctor. Burnout at work can be another source of fatigue. Symptoms such as reduced job performance and productivity, lack of creativity, or low energy may show burnout. Even if one isn't working, a lack of meaningful activities during the day can lead to similar experiences and fatigue.

The first goal of this module is to increase your awareness of the various contexts where you experience fatigue. Mindfulness is a way of strengthening your ability to notice your present moment experience. That can include your environment, as well as your inner sensations, thoughts, and emotions. There are a variety of mindfulness exercises within this book. I encourage make mindfulness a daily practice to expand your ability to notice what shows up in each context where you experience fatigue.

We will begin this module with a mindfulness exercise designed to help you realize you are not alone in your struggle. Fatigue can feel very isolating. When other people want to do things, and our mind tells us we're too tired, that we don't have enough energy, the result may be that we get left behind. Being fused too tightly with what our mind tells us can lead us to make choices that distance us from other people, and from doing activities we love. One thing that can be most helpful in confronting fatigue is an awareness of the people around us and the support that we have. I sometimes see my battle with FSHD as a solitary one. I can lose awareness that those around me will support me in my struggle. I try to go it alone and then end up missing out on resources that could’ve been helpful. If you behave similarly, the following mindfulness exercise is one that may help increase your awareness of that support in the present moment. I based this exercise on a similar one published by Walser and Westrup.[3]

EXERCISE 3.4.1 We’re all in this together

I would invite you to begin this exercise by placing your feet flat on the floor and sitting upright in your chair. Find a position that is stable but not rigid. Place your arms in a comfortable position at your sides… This posture helps you stay focused and alert.

So let’s begin by first noticing your environment. Feel yourself sitting in the chair and your feet touching the ground. Close your eyes. Focus your attention on the tip of your nose and feel the air moving in and out as you breathe… Notice your breath.

Now extend your awareness to your body, noticing each of your senses - what you feel, sense, and hear. Be aware of your body’s position and all the sensory experiences it is having.

Now imagine that a part of you could float above this room and see yourself–sitting in the chair from overhead… in your mind, picture how FSHD affects your body. How you sit, how you hold your arms and your facial expression. Observe the unique solitary struggle that you have with this disease.

Allow yourself to rise higher, through the ceiling and above the building. Using your imagination, picture everyone in your country who has FSHD - hundreds, thousands, maybe tens of thousands… Boys, girls, women, men all gathered together in a huge open field - together in one place. Some are in wheelchairs, some have canes or walkers, and some have family members or attendants with them to help. They come in every age, shape, size, color, background. Notice all the effort it took for them to come together, the struggles each one has that are like your struggles. Notice they have all experienced losses and physical decline. They struggle with confidence and self-esteem. Some of them are worried or sad or lonely or angry. Others are hopeful, with smiling faces, filled with joy. Notice you are together with them, sharing the same struggles.

Picture yourself floating higher and seeing your country's entire population below. Still using your imagination, picture everyone below going about living their lives... struggling with diseases of every sort. Notice them living their lives, having feelings—some good, some bad—having anxiety, having love, growing, changing, hurting, crying, loving, and struggling with unique challenges.

Now allow yourself to float even higher, out of the atmosphere, so that you are floating high above the earth. You can see the entire planet. Now imagine, as best you can, all the billions of people living their lives. They have different cultures, speak different languages, they are all trying to live, with pain, joy, sorrow. They all struggle. (pause for a longer period)

Now imagine that you are floating back toward the earth… It is growing in size. Now you can see your continent come into view… Now float down even further until you see where you live. Then continue on until you see your city… and then closer still until you are hovering right over this building… Bring yourself back to this room—notice yourself still sitting here…

Then bring your awareness back to yourself, your body, your senses, your feelings and thoughts, and just note that we are all- whatever our struggles—we are in this together.

Now picture the room you are in. Notice the sounds of the room, and notice that even if you are alone in this room or in your house at this moment, you are not really alone. When you are ready, open your eyes.

* * *

For most of my adult life, my struggle with FSHD has been a solitary one. I had symptoms of the disease for over 50 years before I met another person with the disease. I hope the exercise we just finished helps you be more aware of others around you who share the same or similar struggles. You are not alone!

This next exercise is not one that you can finish in a few moments. I would encourage you to print out the form found in **Appendix 6** before you begin. I would encourage you to hold on to it and come back to it repeatedly throughout the next week. Each time during the next week that you notice yourself feeling fatigued, take out the form and fill out one row.

One of the important goals for this module is to realize the present moment context in which we notice fatigue. That context includes what's happening in our physical environment around us. It includes our social situation—the people near us and the conversations and relationships that preceded that moment. The context also includes our inner experience, the thoughts, emotions, sensations, and memories that are present in that moment. Much of the time, we live our lives on autopilot. We simply react to our environment, both internal and external. We rarely notice or lack awareness about the context of those present moments. The exercise will take several days, but it is one that you can start right now and then come back to again and again.

EXERCISE 3.4.2 My FSHD fatigue-related experiences

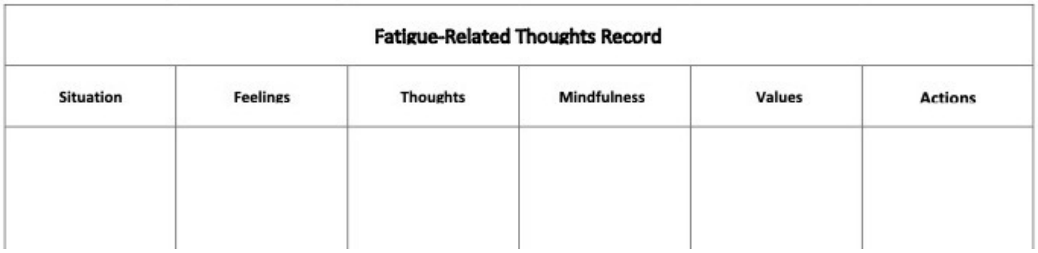
An overwhelming majority of individuals with FSHD report fatigue. However, there are many experiences in situations that produce fatigue. These include:

Physical tiredness, both general and specific. As FSHD progresses and muscles throughout the body weaken, the physical effort required to perform daily tasks increases. Things such as standing or walking or simple chores can leave you feeling exhausted. At other times, the overuse of a specific muscle group can physically exhaust that area of the body. It may cause an overall sense of fatigue. ***Sleep problems*** arise from multiple sources (such as apnea, physical discomfort, worry). Sleep difficulties can also result in tiredness and fatigue throughout the day.

Fatigue may also arise because of ***mental and emotional stress***. Feeling overwhelmed and fatigued can result from daily hassles, conflicts, and worries.

*Symptoms of **anxiety or depression*** can manifest as fatigue, sadness, hopelessness, and discouragement. They can lead to a general sense of malaise and lack of motivation to do anything. Individuals with depression frequently report difficulty, even getting out of bed. We can feel these experiences as intense fatigue.

This exercise increases your awareness of thoughts and feelings when you experience intense fatigue during the week. Examining the context in which you notice fatigue can provide insight into the source of fatigue and allow you to change your relationship with it. Use this exercise to practice mindfulness, acceptance, and value-based actions. Over the next week, choose three to five different situations where you feel unusually fatigued to examine with this form. You will find the form in **Appendix 6** at the end of this book. Print the form before starting the exercise. Be sure to fill in all the information each time.



Fatigue-Related Thoughts Record

Situation	Feelings	Thoughts	Mindfulness	Values	Actions

*In the box labeled **Situation**, briefly describe the fatigue situation. Be sure to note the time of day and where are you are. Think about the people present with you, or if you are alone. If people are present, think about the relationship you have with them and your conversation with them. Think about what was happening just before and during this present moment. What was the emotional tone of the moment? Did you have any urge to do or say anything during the situation?*

*In the box labeled **Feelings**, write all the feelings that you notice in the current moment. Specify emotions (anxiety, apprehension, fear) and physical sensations with locations. These might include tension, or pressure, or even physical pain in some particular area of your body.*

*In the box labeled **Thoughts**, jot down the thoughts going through your mind as you are in the situation. There are several types of thoughts you should pay particular attention to. First, notice any thoughts specific to the present situation, any meanings, or explanations your mind provides. Second, note any thoughts about yourself. These might be beliefs or evaluations and judgments about your character or capabilities. You might notice thoughts about past events or perhaps even worries about the future. Notice whatever shows up in the ongoing narrative of your mind. Make a note of these in the box labeled thoughts.*

Now, take a few moments to engage in mindful awareness exercise. You could consider returning to the Leaves on a Stream exercise found in Part Two. Use the exercise to observe and recognize your thoughts as thoughts and feelings as feelings, as they come into and then leave your awareness. Write out notes about your observations in the box titled ***Mindfulness****.*

Now, take a few moments to reflect on which of your values are most salient in this present moment situation. Write a statement of that value in the box labeled ***Values****.*

In the ***Actions*** *box, try to identify 1-3 specific actions that would express the value you listed. Goals are specific actions that give life to, or move you toward, your most important values. Try to set specific achievable goals that you could do beginning today. Then allocate time and commit to doing them even if you are tired and have no energy.*

* * *

I hope the previous exercise provided some insight into the context surrounding your fatigue on multiple times, days, and places. Hopefully, you have gained an awareness of what your mind says to you at these times. Noticing the emotions present with your fatigue can provide clues as to the cause. Mindfulness allows you to notice the present moment and make choices that reflect your values.

Fatigue often functions as a way of avoiding unpleasantness. Whether that unpleasantness is the physical sensation of tiredness, or a thought telling me I don't have the energy to do anything. By applying the processes of willingness when I have the thought that tells me I'm too tired to do something - I open up space to make a different choice. I can loosen my grip on the firm belief that I cannot act in the presence of fatigue. I can make an intentional choice about what is actually best for me, and the action that will best express my values at that moment.

Research suggests that often what is best for us when we feel fatigue - is that to get up and get moving about. Ideally, you direct that physical activity - toward your values. You will get up and engage in meaningful activities with people you care about. You will often discover over time that social engagement and physical activity energize you. But it is also important to realize our muscles have limits, and not to overwork them, or do things likely to injure you.

The next piece of the fatigue puzzle is having a clear sense of what is important in our lives' most precious areas. Clarifying what our values are amid all the varying demands of life is an important exercise. Below you will find an activity adapted from one created by Dr.Walser and Dr. Westrup.[3]

EXERCISE: 3.4.3 Clarifying my values

Below is a list of several domains of life that most people find to be important. Look through the list - you may find that you have values in each of these domains, or you may find that you have values only in some of them.

This module is focused on fatigue, and data suggests that increasing your level of physical activity is likely to reduce fatigue. As you complete this exercise on values, try to identify specific actions that express these values but also involves more physical activity and engagement with people you care about on your part.

Pick one or more of the domains that are important to you. This exercise is not a test to see if you have the “correct” values. Instead, work on describing the qualities that you would like to live out in each important domain. Describe how you would like to treat people, including yourself, if you had the ideal situation. Elaborate and use additional sheets of paper.

Life Domains/Values:

- Marriage/intimate relations values
- Parenting/child-rearing values
- Extended family relations values
- Friendships/social relations values
- Employment/ Financial values
- Educational/learning values
- Leisure/ recreation/ citizenship values
- Health/ physical well-being values
- Spiritual/religious values

To complete the values form below (*Use extra paper and make more copies to work through* *each selected life domain*):

1. *Pick a life domain where you have an important value. Describe that value as you would describe it if no one else would ever read this worksheet. Be bold.*
2. *Rate the importance of this value to you using the following scale: Low = not at all important; Medium = moderately important; High = very important.*
3. *Describe several specific goals (actions) that could help you live each value. Choose goals you can do regularly and/or immediately.*
4. *Write a thought or emotion that might prevent you from doing a specific goal.*
5. *Write a brief paragraph about what it would mean to you to live the value and what it would mean if you didn't.*

Selected Domain/Value:

Describe the value:

Importance of value: (Low, Med, High)

Goals: (specific doable actions that express the value) Try to identify actions that involve increased physical activity.

1.

2.

3.

Obstacles (Thoughts or emotions that might keep you from acting):

What would it mean for you to live the value?

What would it mean for you NOT to live this value?

Work through each of the life domains above that are relevant for your life. You may find that several of these domains overlap. Do your best to keep them separate. Remember, a value is something that you can always work on—it is your compass direction, not your outcome. This is not about being pragmatic. We want you to explore what's important to you, what you care about most, what you would like to work toward, in the ideal situation, if nothing were impossible. While doing the worksheet, pretend that magic happened, and that anything is possible.

Because this module is focused on fatigue, remember evidence indicates that increasing physical activity often leads to reduced fatigue. In identifying actions express your values - pay special attention to actions that will increase your physical activity and improve your connection to people that you care about.

Once you're finished, take several tangible goals related to one Domain/Value and set aside time in the next 24 hours to get active. Commit and Act—regardless of your present level of fatigue. Take a single forward step—and then another - keep moving!

◆

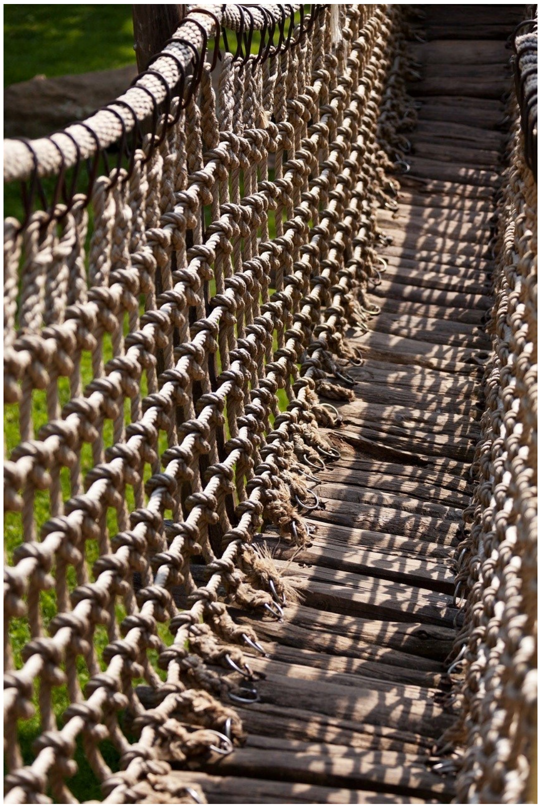
Living a life that matters is not a onetime choice, it's a journey. Having FSHD with fatigue present is also not a single experience. It's also a journey. We are not always aware of it, but every day and every moment, we have the power to pause, choose our direction, and to take action. But it's difficult. It can feel frightening. Sometimes we come to a place where it feels hard to move forward. We are not sure if we can move forward—whether it's safe or scary.

David Gillanders developed an exercise called "The Rope Bridge."[4] To me, it sort of conveys that sense of being stuck and afraid to move forward on our path. It might also help clarify the choices we face when fatigue is present with FSHD.

◆

EXERCISE: 3.4.4 The rope bridge

Imagine you are standing up on a high cliff overlooking a deep, narrow canyon. Across the canyon, you can see something you can just make out - it is a glimpse of that special place you've been looking for. It's a place where you can live vibrantly, where life would feel full and exciting. It seems like a place where friends and family could gather with you, where you could feel delighted. And in front of you - you can see that there's a way to get to the other side of the canyon. It's an old rope bridge. But, as you look at it, you're not sure the bridge is safe. It seems like there might be a few planks missing. Maybe the ropes look a bit worn. But you are not sure that it isn't safe either. And there, on the other side, is the place where you would like to be. If this were the situation, what choices would you have?



Rope Bridge

Take some time to process this. You could decide the bridge is definitely unsafe and that you are reluctant to take the actual risk involved in moving forward in that direction. You could also decide to just go for it and run across the bridge, ignoring the risks. That's a brilliant choice unless the bridge is dangerous and will cause injury. Taking actions that express our values always requires at least a small leap of faith because we don't always know what the outcome will be. We take risks - not always physical, sometimes emotional or relational in the service of doing something that matters. The third option would be to move forward cautiously, one step at a time, ready to turn back if it is impossible. The exercise is a metaphor to show that you can't determine the safety of what lies ahead by standing still. To continually live our values, we will be required to risk stepping into an uncertain future. Not every action we take will work out as planned, but by persevering, we will keep moving in our chosen direction.

This is an experience I often have with my FSHD as it continues to progress. There are activities I want to engage in, but I also need to ensure that I can take part without injury. I may need to adapt in certain ways to my changing body. My mind sometimes overestimates and, at other times, underestimates the actual risks. It's important to consult with other people to get a clearer perspective. What I am trying to do, however, is to continue to make physical activity my default. Daily, I try to make physical activity a priority despite any conflicting thoughts or emotions..

* * *

This rope bridge metaphor is quite similar to what we experience with FSHD and fatigue. Research tells us that exercise and activity reduce fatigue. Yet, our minds often tell us it's impossible, that we don’t have the strength to do it. The only way to determine what is right is to keep moving forward and see what happens. And in order to do that, we have to be willing to experience emotions of hopelessness and discouragement, and thoughts telling us it won’t work. If we can hold those experiences lightly, allowing them to be there - we can choose to take steps toward the things that matter to us.

Fatigue seems to be nearly universal for people with FSHD. Research shows that effective strategies to reduce fatigue are counterintuitive. Exercise helps reduce fatigue for people with FSHD, despite feeling too tired to do it. It can be a challenge to find a workable balance point in physical activity between too little and too much. It requires releasing the grip that thoughts like ‘I’m too tired’ have on us and engaging in healthy physical activity. Simultaneously, we must monitor our bodies to ensure that we don’t overtax certain muscles and injure ourselves.

In this module, we explored the role that fatigue plays in the lives of people with FSHD. We engaged in a mindfulness exercise designed to enhance our awareness of our shared struggle together. Those with FSHD are not alone - we can support one another on our journey. We also explored some of the fatigue-related thoughts that show up, and the context in which those thoughts emerge. We looked at important life domains and explored the values that we want to stand for in those parts of life.

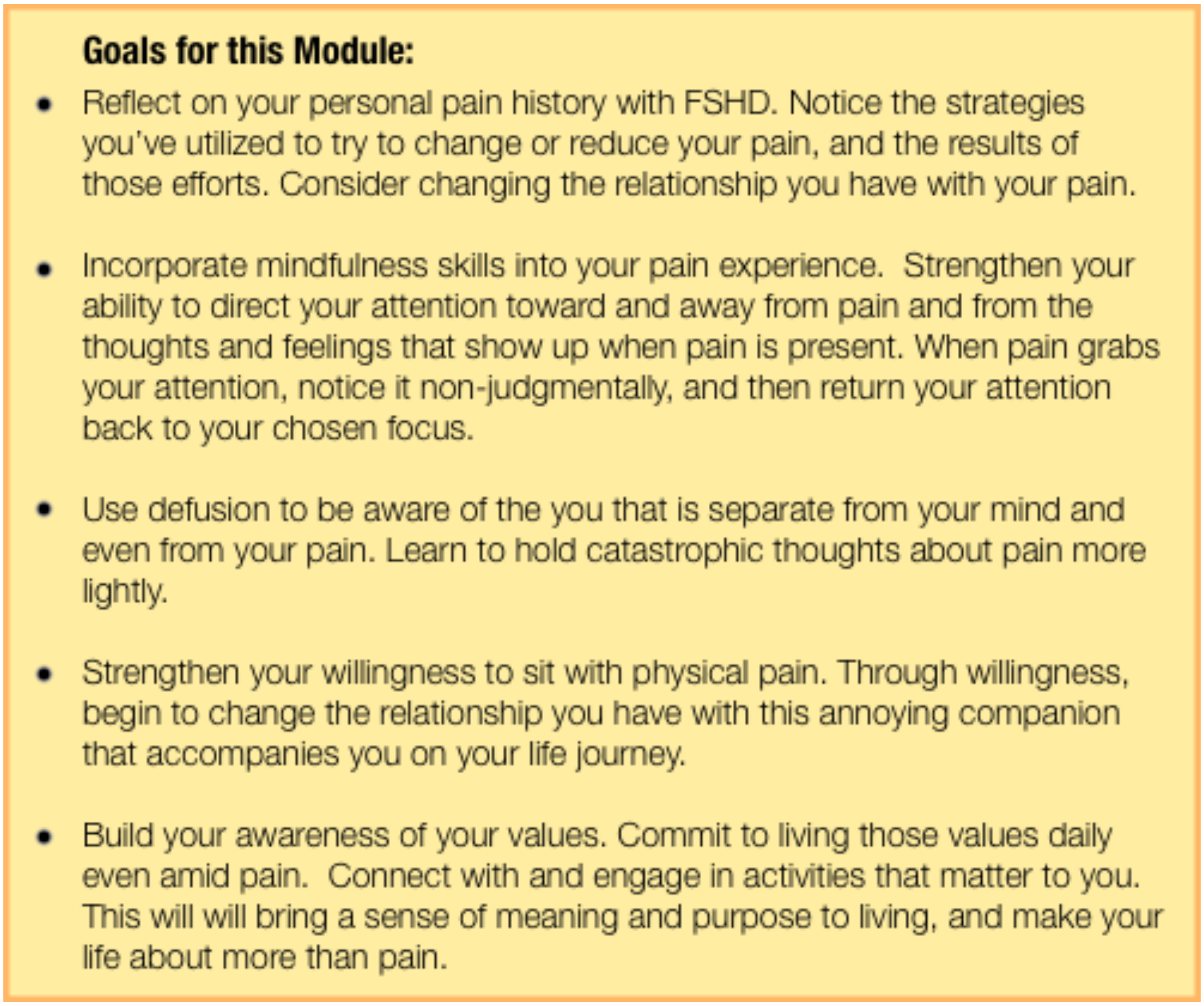
In the next module, we address another common FSHD symptom - chronic pain. As we noted in this module, pain and fatigue are connected. Chronic pain plays a role in increasing fatigue. This next module will explore the mental and emotional context of chronic pain in more detail.

MODULE FIVE

Managing chronic FSHD-related pain

Changing our relationship with our bodies and chronic pain caused by FSHD progression is the focus of this module. Looking beyond FSHD pain, chronic pain is a major issue that affects over 20% of the general population. Studies show that there is a bi-directional relationship between chronic pain and mental health.[1] As mentioned in Part One, individuals with chronic pain have a much higher prevalence of mental health problems. Individuals with mental health problems are also much more likely to experience pain. Evidence suggests this two-way link between pain and mental health also exists in people with neuromuscular disease.

The American Psychological Association (APA) recognizes ACT as an effective therapy for both chronic pain and mental health issues. If chronic pain is a significant issue with your experience of FSHD, I hope you will work through this module carefully.



Goals for this Module:

- Reflect on your personal pain history with FSHD. Notice the strategies you've utilized to try to change or reduce your pain, and the results of those efforts. Consider changing the relationship you have with your pain.
- Incorporate mindfulness skills into your pain experience. Strengthen your ability to direct your attention toward and away from pain and from the thoughts and feelings that show up when pain is present. When pain grabs your attention, notice it non-judgmentally, and then return your attention back to your chosen focus.
- Use defusion to be aware of the you that is separate from your mind and even from your pain. Learn to hold catastrophic thoughts about pain more lightly.
- Strengthen your willingness to sit with physical pain. Through willingness, begin to change the relationship you have with this annoying companion that accompanies you on your life journey.
- Build your awareness of your values. Commit to living those values daily even amid pain. Connect with and engage in activities that matter to you. This will will bring a sense of meaning and purpose to living, and make your life about more than pain.

"When suffering knocks at your door and you say there is no seat for him, he tells you not to worry because he has brought his own stool."
— Chinua Achebe

My FSHD Pain Experience

When I was first diagnosed with muscular dystrophy, I was told by my physician that "*painless weakness*" characterized the disease. I've since learned that for many people with FSHD, that characterization of the disease was completely wrong. For nearly all of us, pain will be at least an occasional burden on our FSHD journey. For some, however, pain will be an overwhelming and daily part of that journey. I am very fortunate that so far my experience of FSHD has been mostly pain-free. I've had occasional, sometimes mysterious, muscle pain over the years. Usually it passes quickly. Only a few times has my pain become chronic (lasting over three months). Physical therapy or massage helped to relieve the pain each time so far. However, these occurrences are becoming more frequent, and I don't know what my future holds. I've learned through social media posts and from face-to-face meetings with other FSHD patients that I have so far been quite lucky. Pain, sometimes excruciating and enduring, is a part of the experience of many individuals with FSHD.

As mentioned in Part One, results from the PRISM-FSHD study showed that 82% of individuals with FSHD report significant pain. This result is consistent across age groups and is uncorrelated with duration of symptoms.[2] Nearly 30% of FSHD patients report severe chronic pain (often shoulders, lower back). This pain impairs physical functioning and quality of life.[3] Pain from FSHD can result from many sources.[4] Overuse of disease-weakened muscles may cause injury and pain. Significant pain in the shoulder or back can be caused by weak muscles that allow partial joint dislocations or create postural or skeletal problems. Changes in gait (walking) may result from asymmetrical weakness, where one side of the body is weaker than the other. This may strain certain muscle groups and contribute to joint pain.

How best to help individuals experiencing chronic pain has been controversial over the last several decades in the US. In recent years, media attention has made the U.S. public aware of the ”opioid crisis.” Rapidly escalating deaths from opioid overdoses over the last 15 years have made the news. We live in a culture that expects that taking a pill will ease suffering of all kinds. Commercial advertisements bombard consumers, prompting them “*talk to your doctor.*” TV ads promote the newest medication for seemingly every disease under the sun. Avoiding physical and mental discomfort of every kind through pharmaceuticals is a gigantic business.

Physical pain is an enormous, complex, and confusing subject. Research suggests that there are several types of pain. Some pain has an obvious source - such as a cut, a burn, a twisted ankle, broken bone, or a fall onto a hard surface. Nerves just under the skin's surface provide sensory information to the brain that it interprets as pain. The brain instructs muscles to protect the body during intense pain, such as pulling away from a hot stove. Other pain stems from internal disease processes such as cancer. Here, pain may be more diffuse and hard to isolate. One may feel it in locations far from the true source.

For this module, the most important distinction in understanding the impact of pain is between acute pain and chronic pain. Acute pain originates from a specific injury or damage to tissue that, in most people, heals and recovers. When pain persists for 3 to 6 months after an injury, experts consider it chronic. Research shows that when pain continues to exist for three months - there is an 80% chance, it will persist four years later. That is because chronic pain that continues after an acute injury has healed is likely very similar to a traumatic event's memory.

Some researchers have described chronic pain as a persisting aversive memory network. This is absolutely not to say chronic pain is not real or that it's all in your head. The pain is genuine - it can continue to affect your life choices and behavior, as well as your satisfaction with and quality of life. Amputees provide an extreme example of chronic pain flowing from a memory network. Nearly 85% of those who have a limb removed still feel pain in the limb long after it's gone. One of the other challenges of chronic pain is that pain medications are much less effective. This can be extremely frustrating and even dangerous for patients, as there is a tendency to overuse medications or go to extreme lengths to stop the pain.

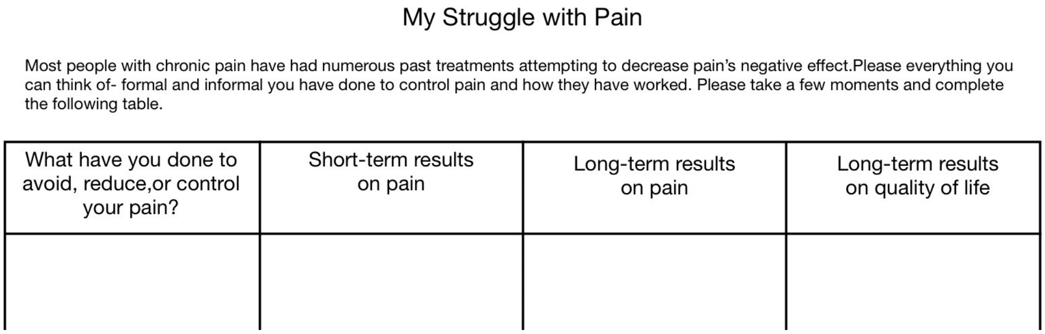
Research shows that psychological processes profoundly affect the impact of chronic pain. Chronic pain severity and functional impairment are related to psychological and social factors. Individuals with the same disease process will often vary in how they report the severity of their pain, and how they respond to that pain.[5]

The goal of ACT in this module is to change the relationship that we have with chronic pain. We seek to change our relationship with the thoughts and emotions show up during pain. We want to open ourselves to living a more active, meaningful life, even when pain is present. The first exercise in this module focuses on exploring your personal experience of and responses to pain. I slightly changed this exercise from one found in a chronic pain treatment guide written by Lance McCraken.[6]

EXERCISE 3.5.1 My Pain History

The goal of this exercise is for you to reflect honestly on your own personal experience with pain. I would ask you to set aside significant time without interruption to engage in this exercise. Below you see a small sample image of part of a worksheet that you will find in

Appendix 7. Print out a copy of the worksheet.



My Struggle with Pain

Most people with chronic pain have had numerous past treatments attempting to decrease pain's negative effect.Please everything you can think of- formal and informal you have done to control pain and how they have worked. Please take a few moments and complete the following table.

What have you done to avoid, reduce,or control your pain?	Short-term results on pain	Long-term results on pain	Long-term results on quality of life

Now, I ask you to just sit for a few minutes and consider how long pain related to FSHD has been present for you. Notice whatever thoughts, feelings, or memories arise as you reflect on your experience with FSHD pain. Now, fill out the worksheet, starting with the left-hand column. Try to make an exhaustive list of everything that you've ever done over time to avoid, reduce, or control your pain. You might have coped by taking medicine, or using assistive devices. Perhaps you limited movement, saw doctors, exercised, or avoided exercise. Maybe you changed jobs or stopped working. Try to list everything that you've done to reduce, or keep from developing, pain as your FSHD has progressed. Once you feel you've listed everything - go back through the items on the list one at a time and complete the next three columns of the worksheet. What was the immediate impact of each list item on your pain? What was the long-term impact on your pain? Finally, did engaging in the activity on the list ultimately over the long term improve the quality of your life?

There are no right answers here. This is about your lived experience - no one else's. Reflect honestly about what has happened as you tried to manage your pain. Take some time to think about the following questions.

- Does your experience show that you can reduce your FSHD pain? In the short term? What about over the long term?
- Does your experience show that the actions you take to reduce pain improve your life? Do your efforts make your life larger and more free? Or do those actions make your life smaller, more restricted, and more dominated by pain?
- Do you have any sense that the solutions you've tried have become part of the problem? Is there a 'vicious cycle' quality to what is happening with you related to pain?
- Recognizing some things do decrease pain - what is the ultimate outcome of your

> efforts? Does your life revolve around eliminating pain or is it focussed on living fully?.

* * *

Perhaps the quote attributed to Henry Ford applies if any of these are the case. *“If you always do what you’ve always done, you’ll always get what you’ve always got.”* If it feels like you’ve been spinning your wheels with chronic pain - perhaps it’s time to consider trying something new. Why not try to develop a new relationship with your FSHD pain?

There is a metaphor used within the ACT Community that's highly relevant to the individual struggle people with FSHD have with chronic pain. It's called "tug-of-war with a monster.”[7] It's a metaphor that helps to make sense of the ACT practice of willingness. The metaphor is as follows.

> *Imagine that you're standing on a path engaged in a life or death struggle—an actual tug-of-war with a huge hairy, ugly, screaming monster. You’re locked in this battle. Each of you holding onto one end of a strong rope. You pull and pull and pull to get away, but escape is impossible. The harder you pull one way, the monster pulls back to even harder. You’re locked in this battle with no end in sight, at risk of falling into a chasm at every moment. There's a path leading away from the cliff, but you can’t move down that path because you’re trapped in the battle. The louder you scream to be free, the louder the monster yells back in rage.*
>
> *So what do you do if you’re locked into a terrible battle with a horrible monster? The answer seems simple—Drop the rope! What happens when you do that? When you drop the rope, you turn away. You realize you can move again. So you walk down the road, but you find the monster follows you. He’s right beside you. He’s still raging and screaming at you, and trying to stop you from moving. He’s trying to hand you back the rope so you can continue your battle. What actually changes when you drop the rope? The monster doesn’t change - he is still yelling and screaming and trying to get you to battle with him. What changes is that without holding onto the rope, you can move again. You can move in the direction you want, despite the monster's screams beside you. Your power is not the power to make the monster go away. Your power is the ability to choose to go in the direction you want to go. But only if you’re willing to let loose of the rope, move your feet, and allow the monster to continue to be loud and scary.*

Chronic pain in FSHD can lead to negative feelings like frustration, discouragement, hopelessness, and despair. It may also bring thoughts that tell us we are powerless and will never have the life we want. Our one freedom in all of this is our ability to choose to continue to move toward what matters to us. We may not be able to make pain go away, nor can we make our mind stop telling us it's hopeless, or even push away our painful emotions. What we can do is choose to take action. We can just loosen our grip on the things holding us back and start putting one foot in front of the other. Movement forward can happen if we are willing. We can move toward our values even in the presence of pain. We just have to let the other stuff come along for the ride.

Given that the module's theme is FSHD-related pain, if you're experiencing pain right now, I'd invite you to use that pain as a focus for this next exercise. But, suppose pain isn't as part of your experience right now. In that case, I invite you to identify some other internal experience that's uncomfortable or that you might wish would go away. It might be an emotion such as anger, sadness, anxiety, or fear. It might be a particular evaluative thought that frequently comes to mind related to your FSHD. See if there's some location in your body where you experience that emotion or thought. If pain is present, identify the location in your body where you feel the pain most intensely.

One of the enduring challenges of chronic pain is that it grabs the complete focus of our attention. This can make it hard to focus on other things, or to decide. Pain often causes our minds to say "*I can't do that*" or "*I don't want to do anything*." This module is not advocating ignoring pain, or not using appropriate pain medications when necessary. The module's goal is to help us refocus, make value-based choices, and live a vibrant and meaningful life. Clinging to our thoughts as truth can hinder us from meaningful actions. Defusion from thoughts is one way to loosen the grip of those thoughts on our behavior.

Mindfulness practice helps us intentionally direct and redirect our awareness in the present moment. This skill lets us sit with our present experience and choose our actions. The following exercise provides a way to practice being present to our physical sensations of discomfort or pain. I have changed the exercise from one provided on the Living Well website.

EXERCISE 3.5.3 Mindfulness of Pain

Instructions: *As we have done previously, settle into a chair. Place your feet firmly on the floor, relax your arms at your side or in your lap. Bring your awareness and attention to your* *breathing. Pay curious attention to the sensations as you breathe in through your nostrils, and then as you breathe out again.*

After a couple of minutes, bring your awareness to the physical sensations throughout the rest of your body. Notice what is happening in various parts of your body. Observe places that feel comfortable and those that feel discomfort. Bring your awareness intentionally to some part of your body that feels discomfort. It could be an itch, or tingling numbness, or even pain, whether mild or severe. Notice any urges associated with the area. A desire to scratch, or move, or readjust into a more comfortable position. Allow your awareness to sit with that urge - don't act on it.

Notice the thoughts that show up in your mind as you focus on this uncomfortable sensation. Notice them simply as thoughts that you have. Realize you can choose to act or not act on these thoughts. Whatever the thought might be, just let it occur without acting on it. Continue noticing the thoughts and the feelings and sensations. Notice how they might shift and change. One thought giving way to another thought. Sensation becoming more intense or maybe diminishing.

Now, shift your attention. Scan your body to see if there's another area where discomfort is present. This experience might be more intense or less intense. Repeat the exercise with your awareness of this new discomfort. It might be similar or dissimilar from the area you have focused on previously. Allow yourself to sit with this discomfort without needing to do anything about it.

Continue this exercise as long as feels appropriate. After a few moments sitting with a sensation, shift your attention to another part of your body with a different, uncomfortable sensation. Sit with that. Notice that you can choose to be with your physical sensations. Allow your pain to be there while choosing not to act on the urges and thoughts that show up with the pain. Notice the part of you that can decide whether to take action. Then take action, even in the presence of pain.

Finally, bring your awareness and your attention back to your breath. Notice your breathing. Pay curious attention to each breath as it flows into your lungs and returns out as you exhale. Mindfully notice the physical sensations as you breathe. After a few moments, return your attention back to the room.

* * *

Mindfulness is a skill that you must develop and practice regularly in order to be easily available and useful in our lives. Mindfulness can play an important role in changing how we engage with the physical pain that sometimes accompanies FSHD. It opens a window into our inner experience of physical sensation, thoughts, emotions, and urges to act. Practicing mindfulness regularly helps us to stop living on autopilot. We live more consistently and intentionally in the present moment. I encourage you to pursue mindfulness, and to use all the mindfulness exercises in this book, as well as the many resources available elsewhere.

I adapted this next exercise from a widely used one shared by Steven Hayes in his recent book—*A Liberated Mind*.[8]

EXERCISE 3.5.3 Doing the Opposite

Instructions: This exercise disrupts the close association between our thoughts and the part of us that makes choices about how we're going to live. We often live as though all our thoughts are true and helpful, when in actuality, that may not always be the case. Our minds work nonstop 24/7 doing the very things that minds do - thinking, planning, remembering, analyzing, evaluating, and telling us what to do. Those things are usually extremely useful, especially in the world outside of our bodies. But because minds are so useful, we can easily fall into the trap of forgetting that our minds also make mistakes. Our minds sometimes tell us to do things that aren't helpful, or that while they may feel good, make our longer term situation worse.

What I want you to do is very simple and will probably feel silly.

Sit comfortably in your chair, extend your arm and wave your hand back-and-forth like you're greeting a friend. Keep waving your hand while you read this next sentence out loud several times. ***"I CANNOT WAVE MY HAND."*** *Keep waving your hand and continue reading the sentence over and over—at least five or six times.*

Now, let's change it up a little. If you are able and it doesn't put you at risk for falling - stand up. Let's do the whole exercise again. While standing, raise your hand a little and begin waving like you did before. This time repeat over and over, ***"I CANNOT WAVE MY HAND WHILE STANDING UP."*** *Repeat this sentence five or six times while waving your hand like a monarch waving at your subjects.*

When you finish, sit back down and reflect for a few minutes on what came up. What were your thoughts while you were doing this? Note any other reactions.

The point of this exercise is obvious, but hopefully, doing something so silly can call attention to something important. Our minds often tell us things about what we can or cannot do. There may be times in our lives when we are in such pain that our mind will tell us, "*I can't stand this pain another second.*" While clearly, pain isn't pleasant, that statement also is not accurate. In the time that it takes to think that thought, another second has passed, and we've endured the pain successfully. Clinging to what the mind tells us about pain may prevent us from choosing to engage in meaningful activity. Paradoxically, meaningful activity is often what will allow us to shift our attention away from pain into life. Staying fused to these thoughts can keep pain as the central focus of our attention. Loosening our grip on those thoughts frees us to shift focus away from pain, toward something else - something more important.

* * *

I find exercises like the previous one helpful because they are quick, fun, and playful. I can remember them and use them at a moment's notice. For me, they serve as a reminder not to take myself and my struggle with my FSHD so seriously. It's easy to fall into discouragement, and feel overwhelmed when my mind tells me I have no future, and nothing will ever get better. In reality, that's the perfect time to tell my mind 'you're not the boss of me.' I quickly do an exercise. I imagine my mind telling me 'you can't clap your hands' while I clap my hands. And then, I imagine my mind telling me, 'you can't stomp your feet' and I stomp my feet. I know that sounds goofy, but it makes me smile and realize - I am in control and I get to choose what I do every moment.

I have adapted this next exercise for FSHD from one published by Jill Stoddard and Niloofar Afari.[9] The focus of this exercise is on willingness or acceptance. This exercise helps change your relationship with physical pain and the distressing thoughts and feelings that come with it.

EXERCISE 3.5.4 Willingness with our pain

Physical pain is unpleasant. Most of us will do anything we can to avoid or control physical pain. That's natural! Prolonged physical pain can trigger negative thoughts and emotions, compounding the suffering. And when a person becomes fused with thoughts like '*I can't stand this pain another minute*,' or '*I'll do anything to make it stop*,' paired with emotions of hopelessness or despair, tragic consequences can ensue.

Strategies for avoiding or controlling chronic pain can become unworkable and unhelpful. Overusing pain meds or pulling away from friends and other supports can be dangerous.. We can see this clearly in the opioid epidemic that has ravaged the US in recent years.

I know that the idea of changing one's relationship with suffering may seem abstract and confusing. This exercise makes that idea more clear and concrete. I'd like to ask you to make this abstraction more real by doing this exercise. One way to get a sense of what willingness is really like is to represent your physical pain and mental suffering in an object and act willingly with the object. This may sound a little silly, but it can be quite helpful.

Find a stuffed animal, new or old and worn out. Put some bandages on as in the picture below to represent your pain and suffering.

Stuffed Bear

Treat this stuffed animal as a welcome companion for the next week, and don't try to dismiss the pain and suffering it represents. There are many ways that you could do that.

- *Carry it around with you and keep it close by when you're working, driving, or eating meals. You can set it in a chair or on a table next to you, or simply hold it in your lap.*
- *Interact with it occasionally in a compassionate, caring way. Cradle it like you would a*

child. Stroke it gently as you would a beloved pet. Say out loud, 'Welcome pain and suffering.'

- *Mindfully pay attention to any urges to fix, change, or move your stuffed animal during the week. Hold it in your hand occasionally and just notice that it's separate from you and is not all of you, just like your pain and mental suffering.*
- *Notice that your awareness of it comes and goes, just like your awareness of your pain. Notice that you can let it just be there, or you can focus on it intently, just like your pain.*
- *Treat your stuffed animal like an invited guest. You might even say something like, 'Come on in, suffering - have a seat.' Remember, you don't have to enjoy suffering to welcome it. Wherever you keep it, give it space to be itself.*

Remember that being present is not something you do to achieve a goal or a particular outcome. Being present is simply being open to whatever shows up in the moment. All we're doing in this exercise is practicing being with our thoughts, feelings, and physical sensations differently. Notice that we're not trying to change the experience; rather, we're trying to change our relationship with our physical pain and mental suffering. We practice being present with our thoughts, feelings, and sensations while ***still doing what matters*** *most.*

* * *

When physical pain becomes our focus, it isolates us, and keeps us from being involved in the things that make life feel worth living. When pain is chronic, and we've had this experience for a long time, we can lose touch with how much of what's important in life we give up. We can lose awareness of our values, which were once so important to us. They have drifted away from our awareness because of physical pain. I designed this next exercise to monitor the role that lived-values play in your life day-to-day and moment to moment. This exercise is creating a check-in and check-out log sheet each day of the week. Starting the day by setting achievable actions based on important values promotes valued living. Setting aside time at the end of the day to see what you have accomplished and to reflect on the possibilities for the next day is a way of continuing progress toward actually living the life you want to live. The next exercise provides a way for you to explore what living a values-based life might look like.

EXERCISE 3.5.5 Lived Values Log

Instructions: *Please complete the following log sheet every day for one week. As you begin, try to identify an important value that FSHD-related pain has made challenging to live. Briefly answer each question. At the start of each day, fill out the first three boxes. Later, complete the last two boxes. Each day, see if there is a unique value you wish to focus on or different actions you can take to express the same value.*

Early in the Day	
List One Important Value You Intend to Live Today in the Presence of Pain	
Specific Actions that Express this Value that You Can Do Today	**Potential Obstacles to Expressing Value** (Thoughts, memories, and/or emotions)

Late in the Day	
Did you complete these Actions? **What Was the Outcome?**	**What Reactions did you Experience to Living or Not Living this Value?**

Reflect on the experience at the end of the week and consider continuing certain actions more regularly.

◆

In this module, we focused on living with chronic pain with FSHD. We briefly explored our history of pain with FSHD, along with the thoughts and emotions that have come along with it. We have playfully engaged with those thoughts. We attempted to strengthen a willingness to move toward our values while allowing pain to come along as our companion. Finally, we have continued to focus on what's most important in our lives - our values. We have taken time to identify tangible activities we can engage in that move us in our chosen direction - toward our values.

This is a brief module. For additional information and support, there are at least two books on ACT and chronic pain you might consider. One book, written by JoAnne Dahl and Tobias Lundgren is titled: "*Living Beyond Your Pain: Using Acceptance and Commitment Therapy to Ease Chronic Pain.*" A second book written by physical therapist Joe Tatta is titled: "Radical Relief: A guide to overcome chronic pain."

The next module is one that is highly personal for me. It has been a focus of mine throughout the writing of this book, and that focus will continue for years to come. The next module focuses on managing body weight and improving health with FSHD.

MODULE SIX

Weight management for FSHD

This module focuses on the challenges of maintaining a healthy weight for individuals with FSHD. As mentioned previously, suffering from FSHD does not protect us from the life challenges that affect every human being. Weight management is a challenge nationally within the US. Current CDC Data suggest that as of 2018, 42% of the US population are obese, with 9.2% meeting the classification for severe obesity.[1] These rates have climbed significantly over the last two decades. From 2002 to 2018, the obesity rate rose from 30.5% to 42.4%. The rate of severe obesity nearly doubled from 4.7% to 9.2% of the U.S. population.

Obesity is linked to life-threatening diseases like hypertension, heart disease, diabetes, and cancer. As within the general population, not everyone with FSHD struggles with too much weight. In fact, some individuals with FSHD are so rail-thin because of the loss of lean muscle tissue that they are at risk in other ways. Even if that is true for you, some of these exercises may be beneficial for you.

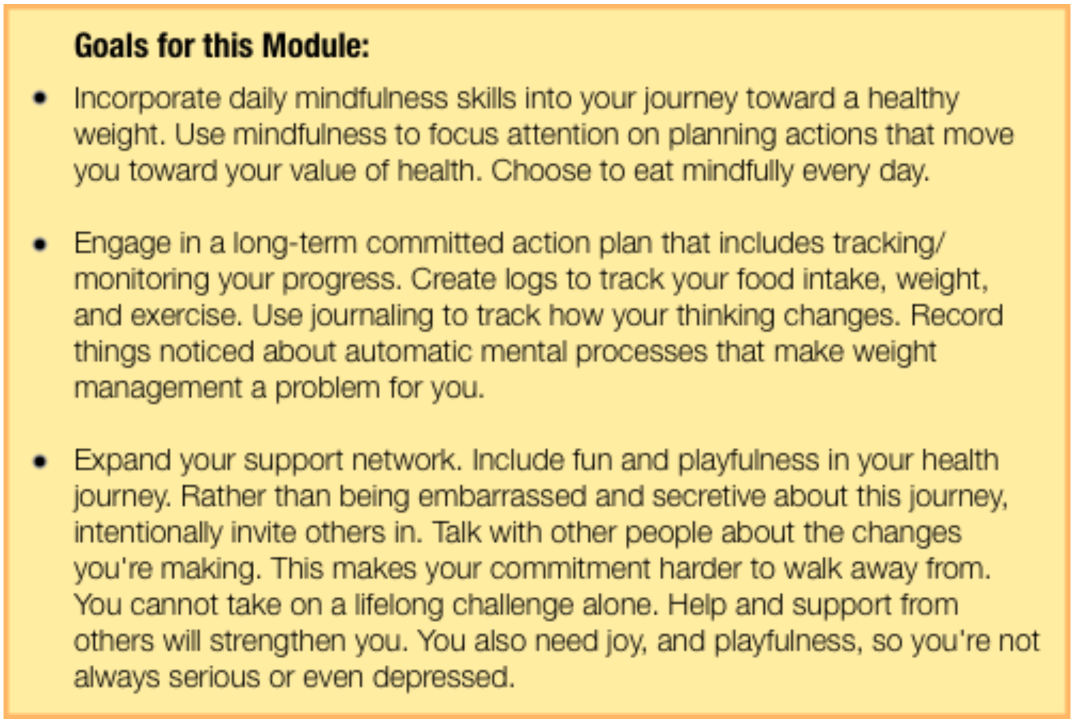
Obesity for an individual with FSHD carries additional risks beyond those experienced by those without the disease. Sarcopenia is a "*syndrome characterized by progressive and* ***generalized loss of skeletal muscle mass and strength*** *with a risk of adverse outcomes such as physical disability, poor quality of life and death*."[2] Sarcopenia is most often associated with aging. However, damage to lean muscle and the corresponding weakness from FSHD - separate from aging, can lead to sarcopenia.

Sarcopenia occurs when a significant loss of lean muscle mass and strength combines with obesity, as is happening more frequently among older adults. This syndrome brings significant risks of physical impairment and disability. Having sarcopenia puts men at eight times greater risk of physical disabilities, including balance problems and falls. These risks are even higher for women, with an eleven-fold risk of having multiple physical disabilities.[3]

There are extra risks of disability and life impairment with this form of obesity. A recent study documented sarcopenia among individuals with FSHD.[4]

Muscle loss, strength reduction, and obesity are risky for the physical functioning and lifespan of FSHD patients. Using every available means to manage weight should be a priority for everyone suffering from FSHD.

Goals for Module 6



Goals for this Module:

- Incorporate daily mindfulness skills into your journey toward a healthy weight. Use mindfulness to focus attention on planning actions that move you toward your value of health. Choose to eat mindfully every day.
- Engage in a long-term committed action plan that includes tracking/monitoring your progress. Create logs to track your food intake, weight, and exercise. Use journaling to track how your thinking changes. Record things noticed about automatic mental processes that make weight management a problem for you.
- Expand your support network. Include fun and playfulness in your health journey. Rather than being embarrassed and secretive about this journey, intentionally invite others in. Talk with other people about the changes you're making. This makes your commitment harder to walk away from. You cannot take on a lifelong challenge alone. Help and support from others will strengthen you. You also need joy, and playfulness, so you're not always serious or even depressed.

My awareness of weight has been lifelong. My mother was very heavy most of her adult life. Involvement in sports helped keep me fit during high school and college and the early years of marriage. My weight remained relatively stable over the next decade and a half, even after returning to graduate school for my Ph.D. My final year of graduate school was challenging. We suddenly had to deal with my parents' medical crisis. We had to move back to the San Francisco Bay Area. I had to get a new job at the VA, and take on a caregiving role. The next several years were a chaotic time of grief and loss; my parent's deaths, financial and family stress, and uncertainty about the future. Looking back, that entire period feels like a blur, but clearly, stress-related eating was a part of it. I put on over 40 pounds.

Near the end of my Ph.D. internship, my supervisor—a young psychologist confronted me. I don't remember his exact words, but he brought me up short during a supervision session. He said if I wanted to succeed as a psychologist, I would need to take better care of myself. I should get my weight under control. Internally, his words offended me. I went home that day, feeling angry, hurt, and humiliated and ashamed.

Stigma about weight

I shared a definition of stigma related to disability in Module Two. Stigma is a negative social attitude held about a group of people who are regarded as deficient in some way. Many groups of people who are viewed as different from the norm (based on race, disability, religion, mental illness etc.) are stigmatized. Obesity is a characteristic that stigmatizing. Stigma of being overweight can't be hidden because it is physically visible. Society thinks that weight can be controlled, and sees overweight people as weak, selfish, and sometimes even immoral. These negative attitudes are widespread. We learn these attitudes early in life, and it is socially acceptable to hold them, as opposed to some other kinds of stigma. When stigmatizing beliefs are acted upon, discrimination results. Weight-related discrimination affects employment, education, ability to find housing. Societal stigma affects relationships and activities with others.[5] Stigma that is directed inward (self-stigma) leads to self-devaluation. In **Appendix 9**, you'll find a copy of the Weight Self-Stigma Questionnaire (WSSQ), along with scoring instructions.[5] The 12-item scale measures both enacted stigma (discrimination), and self-devaluation. If you have struggled personally with weight, I would encourage you to complete the scale prior to beginning the exercises in this module. After completing the module, wait a few weeks or months and complete it again. Compare your scores.

Jason Lillis and colleagues used ACT to reduce weight-related stigmatizing thoughts and increase psychological flexibility.[6] They provided no instruction on weight loss strategies to participants. Even without targeting weight loss specifically, the study found that participants had improved quality of life, less psychological distress, and reduced BMI at the three-month follow-up.

I know I learned stigma related to overweight growing up, and those attitudes have affected my weight management efforts over the years. It's been about 30 years since that day my supervisor confronted me, and my weight and the meanings attached to it have never been far from my thoughts. Twice during those nearly 3 decades, I marshaled all of my mental and physical resources. I engaged in purposeful weight loss activities that led to my losing 80 pounds the first time and 70 pounds the second. I achieved it twice by carefully tracking my food, being intentional about what and how much I ate, and exercising intensely every day. Each time, I kept the weight off for about a year, but then slowly, my weight began creeping back until I regained all the lost weight.

Earlier, we defined experiential avoidance as *"attempts to avoid thoughts, feelings, memories, physical sensations, and other internal experiences—even when doing so creates harm in the long-run."*[1] In reflecting on how that happened, there are two observations that stand out to me. In both cases, it ultimately was **feedback from outside** that brought me to the tipping point where I could choose to change my behavior. But why did it take a dramatic life event to bring me to that point? Why, after losing the weight, did I lose focus and regain it? I now believe that in each case, I had, for years, systematically **avoided** the daily feedback that was right there in front of me.

One way to think about experiential avoidance is to see it as **turning away from feedback**. Feedback made me feel bad about myself, so I avoided to it. During the years of weight gain that led to both previous tipping points, there was ample feedback available in my world. But I would see the bathroom scale and turn away, unwilling to step onto it. I would glance in the mirror but quickly look away again. I would carry the camera and **take** the family photos so as not to **be in** the photos. I would try not to notice that my belt and my waist size were once much smaller.

Here's the thing, though. Experiential avoidance is bigger than just those things. Those things are the surface level. They are the automatic small choices made every day—without even consciously thinking. I do them so frequently, with so little awareness that they become a part of who and how I am. They are automatic. But **why** do we do those things? What drives us to make choices that contradict our values and keep us from the life we want? These are the intense emotions, the painful memories, the difficult, ugly thoughts that are too uncomfortable to even be around.

Emotions pair with equally disturbing thoughts. "*You're ugly,*" "*You're weak,*" "*You can't change,*" "*You're a failure,*" "*It's too late to change now,*" and many, many others. Struggling to not have those uncomfortable thoughts and emotions motivates my avoidance.

I avoided dealing with my problems and made unhealthy choices until some big event made me change. Wouldn't it be better if I strengthened my ability to focus, and to be persistently intentional about the things that I do (and eat). This would build a lifestyle that expresses my highest values and keeps me alive long enough to see all those values fully expressed within my family.

In 2019, medical feedback again alerted me to the need to begin better managing my physical health. This time, Since that time I've used the psychological tools I've taught my clients on myself.

I would invite you to think for a moment about your own experiences or struggles with weight. Take a moment to think through some of these questions.

How have life experiences affected your weight? What painful thoughts and emotions are present for you? Has avoidance played a role in struggles you may have had? What efforts have you engaged in to control or manage your weight? Is your body weight connected to the progression of your FSHD? Aside from FSHD, to what degree is your weight connected with other aspects of your physical health? What are your values that relate to physical health and FSHD?

The first exercise in this module will extend the mindful breathing exercise that we did in Part Two. The activity focuses your attention on the health-related value that is a part of your efforts to maintain a healthy weight. I use a shortened version of this exercise at night just before going to sleep, or more often, the first thing when I wake up in the morning. I find it useful to focus on the values that motivate my decisions and actions before starting a new day. In those brief moments, I set objectives related to what and how I eat, as well as when and how long that I'll exercise that day. I reflect and then make intentional decisions before I get up.This helps me avoid falling into old patterns of living on autopilot. Taking a moment to focus on my values and choosing specific actions helps me prepare for the day.

EXERCISE 3.6.1 Attending to Values

You may start this exercise by getting comfortable wherever you are, sitting up or lying down. You may wish to close your eyes to limit distractions. Now, focus your attention on the end of your nose or on your chest and begin breathing slowly. Notice the feeling of air in your nostrils, or the expansion of your chest as you breathe in and out. You might notice that the air coming in is cooler than the air moving out from your nose… or you might notice some tension in your chest as your lungs expand outward, and then relax again… Let yourself observe your breath, paying attention to the air as it flows in and out… If at some point thoughts arise, just take a *moment to notice the content of the thoughts, where your mind took you, and then, without evaluating, release the thought and return your focus to your breathing.*

Now, after a moment, turn your attention to the value of health. What does pursuing health look like for you? Perhaps it might mean taking actions that reduce stress or conflict in your life or relationships. It might mean buying, preparing, and eating healthy food. It might require exercising your body to maintain and improve your strength and fitness. Maybe it will mean spending more time with the people you care about, strengthening relationships, and giving or receiving love. Whatever that value might look like for you, focus your attention on it as you breathe in and out.

Once you have that value clear in your mind, choose actions you intend to do today that will express that value and move you in the right direction. Note that choice in your mind and commit yourself to set aside a specific time today to complete that action. Once your decision is in place and your commitment made, return to noticing your breath. Breathe in and out for a moment, and when you are ready, open your eyes and return your focus to the room.

Mindfulness can help us make and follow through with committed actions that move us toward our most important values. Come back to this mindfulness exercise often to remind yourself of what's important and commit to actions that reflect your values.

* * *

This next exercise is something I've done twice during my life when I was attempting to make a lifestyle change, leading to weight loss. My previous journal served as a useful account of my activities and internal struggles over time, which proved helpful. So I invite you to try this with whatever value is important to you and with whatever behavioral goals you set to move you toward that value.

This is not a onetime exercise that you can do in a day. Instead, you will start now and continue over weeks, months, or even years. My current journal goes back almost 2 years at this point - with entries every few days. Doing this will require thoughtfulness and commitment. However, I have found this exercise useful in helping me stay on track with the behavioral commitments to health that I've made. I improved my physical health by taking actions consistent with my values about health.

EXERCISE 3.6.2 My FSHD Health Journal

Step 1: Preparation. *Create a document file on your computer in a location that you can find it. Save the file with a filename that expresses the value you are pursuing (including a start date in the filename). If you're not a computer person, purchase a writing journal to use, and give it a similar title inside the front cover - on the first page.*

Step 2: Write a statement expressing your values. *Write one or two paragraphs describing the specific values you are choosing to support through your actions. Describe ways that trying to avoid or control your thoughts and emotions affected your health. Describe the costs you have paid from these attempts at avoidance and control. Finally, describe each factor that now motivates you to move towards your values despite the thoughts, memories, and emotions that have kept you from acting previously. You can title this section "**The values motivating my life changes**."*

Step 3: Write a list of your behavioral commitments. *Next, make a new section below that paragraph and title it "**My Commitment and Goals.**" Now, I'm going to ask you to make a bullet list of **3 to 5 very specific behavioral commitments** you will make from now forward. Pick actions that will move you closer to your values and desired outcomes when you stick to them. As an example, the following is one of the bullet points I used:*

- I will exercise at least 20-mins daily on the exercise bike at least five days per week, and attempt to increase the intensity and duration of my workouts over time

Step 4: Create a timeline for your target outcomes. *Label a new section as "**My Target Outcomes.**" I would recommend making monthly goals that are achievable with significant effort. For example, in keeping with the previous behavioral commitment example, I might do something like this:*

- By Sunday, **October 17, 2020,** my goal is to have ridden 120 miles on my exercise bike

- By Sunday, **November 21, 2020,** my goal is to have ridden 250 miles on my exercise bike

Finally, I would end the section with a statement similar to the one which is listed below:

These are aggressive targets, the dates are flexible! The important thing is to keep choosing to act daily, persist in those actions, and not give up!

You can go back and revise these target outcomes if the progress is coming more slowly (or more quickly) than you expected. Cross out the dates/ or outcomes and revise. Don't erase the previous targets, though - it's good to compare the original and changed intentions.

Step 5: Journal of what you notice about your inner experience as you go. *Finally, write a new section title and call it something like "**My observations: What I noticed along my journey**." Here, I encourage you to make **at least** a weekly journal entry. Begin each entry with a date so that you can track your experience across time. Entries can be short or long. There are no rules. Your goal is to **write what you notice about your unique experiences on this journey toward change**. Note the times when you feel motivated, encouraged, or, in contrast, moments when you feel discouraged. You might notice specific thoughts or emotions that seem to function as triggers toward giving up, changing your behavior, or going back to old ways. You may notice patterns between events or interactions with others that evoke certain thoughts or emotions. Each journal entry can be a place where you celebrate minor accomplishments or re-commit yourself to staying on track.*

Document the date you achieve each bullet point on your target list. Find some simple way of celebrating your forward movement. Keep this document even after completing your goals.

Continue making journal entries, or even setting new goals. Remember, we can never fully achieve our values. You can always continue to set new goals, to continue your movement toward your values.

Memories of Food

In the outer world - the world where our bodies live if something is bothering us, we can often take control to fix it, discard it, or avoid it. For example, if it bothers me that the house is dark, I can turn on a light. If it's too cold or hot, I can turn on the heat or air conditioning. All of those experiences in our outer world exist in the present moment. We can't, for example, make it warmer or colder last week or tomorrow. Every ability to take action to influence our experience in the outer world happens in the present moment.

Our inner world, however, is quite different. In our minds, we preserve the past in memory. Memory for past events and the ability to bring those events to mind has been a key feature in our species' ability to survive and thrive in the world. Memory for past events can be a soothing gift when we recall precious and treasured moments we had with loved ones now gone. But memory for painful, tragic events or mistakes lives on in our minds as well. Reminders of past events often elicit or “trigger” memories, both painful or happy. For example, suppose something in the present reminds me of a past mistake. In that case, I may experience an onslaught of painful emotion and a cascade of regretful thoughts such as "*Why did I act that way*? " "*What kind of person would do that*?"

What does any of this have to do with weight management? The answer is simple. Our memories, thoughts, and future hopes affect how we eat and exercise now.

A while back, I was sitting in a chair in the afternoon, and my wife and I were discussing dinner. It was hot, and neither she nor I felt very much like cooking. She mentioned the idea of picking up chicken sandwiches and baked potatoes at a local fast-food restaurant. At that moment, I noticed visual images of ice cream popping into my head. An impulsive plan emerged about going through the drive-thru with my wife to pick up the food. My hasty plan included buying a chocolate ice cream cone. Immediately in my head, the visual image of a chocolate cone popped up. I didn’t act on those thoughts, but I noticed the process in particular because it felt so familiar. One way that mindless eating happens for me is through these quick intrusive thoughts and images accompanied by an impulsive plan to eat. I’m not as yet completely aware of what contextual factors elicit these thoughts. I know that my mind holds onto these intrusive images of particular foods. I realize that they often motivate impulsive, automatic behavior that results in eating unhealthily.

Memories of favorite foods can serve as triggers to impulsive behavior. Memories can also serve as triggers to avoid activities that are good for us. I've been riding my exercise bike regularly in recent months. Yesterday was a sweltering day, in the high 90s, even into the late evening. I avoided riding most of the day because of the heat and waited until late in the evening to ride the exercise bike, hoping that the house might cool down, but it didn't.

My ride was hot, sweaty, and kind of miserable. When I got up the next morning, the house had cooled down a lot. My first thought was, I should ride now before it gets hot again. Then my mind remembered how miserable I was last night and my motivation for riding today, even in the cool air, diminished. Learning how to respond to memory triggers can be vital to choosing to engage in healthy behaviors in the present moment. Willingness and defusion are important skills for making healthy choices that align with our most important values. I have adapted the following exercise from one published previously.[7]

EXERCISE 3.6.3 Responding to Cake

One way to avoid external triggers is by literally throwing them away. If you're holding a plate of chocolate cupcakes, you can physically toss them in the trash to get rid of them. Where is the cake now? It's buried in the trash. We've gotten rid of the trigger. What about the emotional connection - the thoughts and cravings? Can you just throw those away? Have you ever tried to do this in the past? Research suggests that when we try not to think about certain things, we actually think about them more. If we can't throw mental experiences away when we don't want them—what can we do? Could we maybe try something different? For this exercise, I'd like you to try something else.

Cake

Take a moment to bring to mind an image of that chocolate cake (or any other food you often crave). Imagine picking it up, taking a bite, and eating it. Then, imagine the flavor of chocolate as it fills your taste buds. Finally, imagine the feel of the texture of the cake on your tongue. Notice any thoughts and sensations and feelings that show up and continue to breathe.

What are you noticing about your body? Is your mouth watering? Can you smell the chocolate? If you feel overwhelmed by sensations, or feel compelled to run out and buy cake, you can always go back to just observing your breath. Remember - you're making a choice to not act on the cravings to eat that arise right now. Just hold them gently and notice what's happening in your body and mind. As you sit with these cravings, see if you can release any struggle you're *having with those cravings and just let them be there.*

Imagine that you can open yourself and make room for the craving for them inside you without having to take any action. Picture your urge to eat cake as a wave and yourself as a surfer riding it with your breath.. Your job is to ride the wave of desire to have and to eat the cake. Notice the urge as it starts and as it grows bigger. Staying with it as it increases to peak intensity. Stay balanced on your board by breathing slowly, while the wave rises and then eventually subsides. You're riding this urge and staying on it rather than being wiped out. Observe your craving with curiosity.

When in the past, have you just sat still with an urge and just observed it instead of reacting? Try that now. Where in your body do you feel it? See if you can picture the areas of your body where you feel the craving. Keep imagining cake. Notice that you can just be present with this image doing nothing about it. Just let the image be present. Try to make room for the craving without having to act on it.

To make cake, you mix flour, eggs, sugar, and cocoa powder and bake it at a certain temperature for a certain time. There is nothing extraordinary about it. It is only inside our mind that cake (or exercise, or anything else) can become fused with our emotions. It then can motivate our choices and behavior, sometimes without our awareness, as we take actions on autopilot.

Experiences inside of our heads are transient. They come into awareness, and then at some point, something else replaces them, unless we hold on to them. Willingness allows whatever thoughts or urges show up in our head. Experience tells us that those thoughts and urges Will eventually be replaced my other thoughts, and the urges will fade away for a time. In the midst of the ebb and flow of thoughts and emotions we can hang on to our one freedom - the freedom to choose what we will DO in the present moment. Willingness exercises strengthen our noticing ability. When we observe what comes into our awareness without responding, we strengthen our ability to make values-based choices when it truly matters.

There's a second part to this exercise that I'd like you to try now. We just explored willingness. Now I'd like you to explore defusion. We will repeat an exercise we did in Part Two using the word milk. Now, we will change it using the word cake. You just experienced how easy it is to take an image of food and bring it to life in your mind by thinking about it. It's sort of amazing how a word like cake comes to life as a mental image that engages all of your senses. It is as if you can see the cake, smell the cake, feel the texture of the cake as though it were real. The word cake can bring with it strong impulsive action urges. But cake is just a word. It has no special power or even meaning apart from what our minds (and impulsive actions) give it. To illustrate, I'd like you to try this next part of the exercise.

Move to a location where you can feel comfortable. I'd like you to say the word CAKE out loud, slowly, over and over, without stopping. Cake,… cake,… cake,… cake… now speed up, continue saying cake over and over faster and faster. Keep going until you can't say cake any faster. Notice what happens. If you are like most people, at some point, the meaning (and the images/ sensations attached) to the word cake fade AWAY. Cake repeated over and over at a very rapid pace sounds like gibberish. The word loses its meaning. We have detached the meaning from those letters, from that sound. We realize the meaning only exists inside our minds, and when that fades, there is no meaning.

Noticing and surfing our cravings for food and being willing to have them without responding is a healthy way of managing our relationship with food. Defusing from the meaning of the words is another. You can practice these skills and make them useful tools.

◆

We can develop skill noticing our inner experience, the memories, thoughts, and emotions that are attached to food. Noticing when these show up can help us be aware of urges toward mindless action that flow from these. Noticing these experiences at the moment they occur provides space to choose actions more consistent with our important values. Doing this repeatedly keeps us moving toward our values. I encourage you to keep this exercise in mind daily. You probably already know the times of day when you are most vulnerable to making unhelpful food choices. In moments when your body signals hunger, see this is an opportunity to practice noticing - and then deciding to respond in a values-consistent way. This can be a useful tool when you are trying to make important behavioral changes.

The next exercise is something that I did spontaneously several years ago when I was engaged in my weight loss journey. I had cut back on sweets, limited my portion sizes, and exercised more during the day. Often at night, just before sleep, I would feel intense hunger. I learned that if I simply noticed the sensation and re-focused my attention on something else, the hunger would slowly fade after a few moments.

Spontaneously, one evening when I felt the sensation of hunger emerge, the 60s song, the ***Sound of Silence*** popped into my head. I don't know why. I could hear the opening line "*Hello Darkness my old friend…*" At that moment, my mind substituted the word *hunger* for the word *darkness*. It became "*Hello hunger, my old friend*."

I revised the song lyrics the next day to emphasize my value of health in my weight loss journey. Over the coming months, each time I noticed the sensation of hunger, this new song came to mind as well. I would smile and remember I was still on track and didn't have to take myself so seriously. Below is the result.

Sound of Silence (*Simon & Garfunkel, 1964*)	***Re-written to convey my new relationship with food***
Hello darkness, my old friend *I've come to talk with you again* *Because a vision softly creeping* *Left its seeds while I was sleeping* *And the vision that was planted in my brain* *Still remains* *Within the sound of silence*	*Hello hunger, my old friend* *I've come to be with you again….* *Because a value gently growing* *Deep within me while I am living* *And the value I have planted in my soul* *Roots and grows* *Within the pangs of hunger*

* * *

EXERCISE 3.6.4 Rewrite a Song

For this exercise, I'd like to challenge you to do something similar to what I did by accident. Think about a song with some history or meaning for you or even a favorite nursery rhyme from childhood. Think about a particular value that you are focusing on at the present time, a value that you want to continue to move toward through your actions. If you aren't sure of all the words to the song or nursery rhyme - you can probably get them from Google.

On the left side of the box below, write the actual lyrics or words to the song or rhyme. Next, devote some time to thinking about ways to change the language to express the particular value you are currently pursuing. There might even be a way to include some actions you hope to take as part of the words/lyrics.

Try to maintain as much of the musical flow as possible so you could sing the song in your head with the new lyrics. Be patient, don't give up too quickly. However, make this playful exercise—a challenge to have fun with - don't take yourself too seriously! If you get stuck, let it sit overnight and try the next day again. Something new might come to you.

Song Title and Lyrics	***Rewritten Lyrics***

When you’re finished, reflect on the experience. And then maybe sing the song out loud—maybe in the shower, if that’s less embarrassing.

My song still comes to mind from time to time when my body signals that I’m hungry. It has become a useful reminder to keep moving forward on my journey toward health. I hope your song will do the same for you.

Throughout this book, we've used mindfulness several ways. Many of the exercises have begun by having you notice your breathing. I had you pay attention to physical sensations, like temperature and chest movement. You've attended to other sensations in your body as well, such as the skin of your forearms coming into contact with the chair. I've had you focus on sound - paying attention to the various sounds that you can hear in the surrounding environment. In this module, I'm going to have you apply your mindfulness skills to the experience of food and to eating/drinking.

One of the stark differences that I recognized early in my relationship with my wife was how our two families interacted around food. In my family growing up, mealtime was most often somewhat ordinary. There wasn't much dinner conversation. We sat together, ate rather quickly, and went on to our other activities. The only time this wasn't the case was on exceptional occasions (often a holiday) when friends or relatives would come over to visit. Those meal experiences were much more animated, with conversation and laughter going on for much longer than usual. Parents would excuse the kids to play together while the adults continued socializing.

What was more usual for us as a family was that eating was simply a practical process of taking in food and then going on to other things. We didn’t take time to appreciate sensations or experience personal closeness. It was more like a daily chore. I realized my wife's family had a different meal experience when we started dating in high school and college. Her family was like my family was on the holidays - every day. Dinner could take three hours, as each family member talked about the highs and lows of their day. During my initial visits, I finished my food quickly and remained silent, while others had just begun eating. It felt a little embarrassing.

Learning to eat mindfully means slowing down the process and observing the sensations food elicits. It means noticing your own inner experiences, and the people and relationships all around you. Eating more slowly and closely observing what I'm actually eating and to the sizes of the portions I've taken makes eating more intentional. It is no longer mindless automatic behavior. When eating becomes intentional, and something I am fully aware of, I have more opportunity to make choices that align with my values.

The exercise that follows is a simple way of applying mindfulness to the experience of eating and drinking and drinking.

EXERCISE 3.6.5 Drinking Tea… or Coffee… or Cocoa

Instructions: Now, we're going to engage in a mindfulness exercise with a cup of tea (or some other hot beverage)

- Boil a pot of water.
- Get a tea bag or a leaf strainer filled with tea leaves and put it into a cup.
- Pour the boiled water over the tea bag or the strainer. Fill the cup.
- Let it steep.

As the tea steeps, sit and closely observe the water change colors. When you first pour the water into the cup, the color will emerge from the tea bag/ball, and the tea will turn light brown (or green, or red). Soon it will grow increasingly darker. Let it continue to steep, and when it is how you like it, remove the tea bag from the water. Look closely at the color. Is there anything you didn't notice about the color before? If so, jot down what you notice.

Tea

Now hold the cup in your hands. Feel the warmth. Have you felt a cup of tea like this before? What memories do you associate with this experience? How does the cup feel? Is it burning hot, or just warm?

Lift the cup close to your lips. Notice the sensation as the warmth touches your face. Blow into the cup and feel the steam rise to your lips. Smell the tea. Take a long slow whiff. Most of your sense of taste is through smell. If you aren't smelling your tea, you really don't taste it.

Now drink a sip. Does it burn your tongue? Is it too hot, or just warm? What does it taste like? How would you describe the taste sensation? Try to note your experiences without evaluating them. Now write about your experience.

* * *

The previous exercise is simply an introduction to the idea of eating and drinking mindfully. The goal for this module would be that you expand your ability to observe mindfully how you eat an entire meal, and the various ways you experience food. I want you to notice how you relate to other people in the presence of food. Mindful eating won't happen overnight. It will take practice and perseverance. Still, it will help you with your goal of weight management. At one level, mindful eating is about your experience of food as you sit at the table in front of your plate. However, mindful eating is more significant than that. I encourage you to notice the physical sensations, thoughts, mental images, impulses toward action that show up when you are in the presence of food. These thoughts and impulses, when unnoticed, motivate automatic behaviors may move us away from our values.

One thing that I've noticed is what happens to me when I simply walk into my kitchen. If I see food sitting on the counter, what reactions do I experience? When I walk into the kitchen, do I have an automatic impulse to open the refrigerator door to look inside? Do I open a cabinet door to see the food stored there? I've learned that I have habits related to food that I do blindly without consciously choosing. Noticing those automatic thoughts and behaviors is the first step toward slowing the process and making it intentional. Eating can become a way of expressing my important values in an intentional and chosen manner. But it takes time. You will need to become a consistent observer first and then make choices to act in ways consistent with your values. Eating food is a necessity. But it can also be a pleasurable, intentional experience shared with others that builds our social connections. We can also learn to eat intentionally while expressing and moving toward our values related to health.

Maintaining Health

As I close this module, I want to write a bit about the thing I have the least confidence about in the future - maintaining my weight. As I mentioned on previous occasions, I lost weight, only to regain it. During Covid and during the writing of this book, I again successfully lost 80 pounds. I am currently below my weight from college. My medical health indicators have improved. I have been able to cut down or even eliminate medications. The strategies in this module helped me do that. Regular (moderate) exercise has helped maintain my strength. Finding healthy foods and recipes that I truly love, and eating smaller portions, has been beneficial. However, importantly, I have continued paying attention to self-monitoring. On January 15, 2021, I achieved my weight loss goal. Since then I continue to I weigh myself 3 to 5 times a week and log the weight in my journal. I track and record my meals, snacks, and exercise. I have celebrated my success. I purchased a compass pendant engraved on the back with the date, pounds lost, and my motivating value. Most days, I wear it under my shirt as a reminder. I have now maintained a relatively stable weight for over two years.

My intention is to keep monitoring, and to notice if things change - and take actions consistent with my values. I am fully confident that you can do these things too.

In this next module, I broaden the focus of the book - to include the impact that FSHD has upon our family and friends. FSHD is a genetic disease that affects multiple family members, even those who do not have or carry it. Whether it's FSHD or other struggles, ACT is potentially helpful for anyone going through a difficult time. I would invite you to continue on into this final module.

MODULE SEVEN

ACT for caregivers and family members

So far, this book has revolved around the individual needs, challenges, and struggles of those who suffer from the ongoing progression of FSHD. However, as a genetic disorder, FSHD shows up within families. It affects the lives of each family member, whether or not they have the disease.

ACT is a trans-diagnostic intervention that is relevant for improving anyone's life struggle, not only people with FSHD. Most of this book has focused on the impact of FSHD on those with the disease. However, these same ACT principles, metaphors, and exercises are also useful for family members. This section will not examine all family issues that might arise, as that would be a book unto itself. In this brief section, I will identify ACT processes and metaphors relevant to a few themes that often emerge within families dealing with FSHD. This section will provide two additional exercises that may be helpful. In other instances, you may refer to exercises already presented in previous modules. Look for ways that these can also benefit family members.

Spouse and partner relationships

In a marriage, when one spouse has FSHD, and the other does not, the disease affects the relationship in varying ways throughout their lives. If the diagnosis has occurred before marriage, it may affect the decision to marry. After, it may affect whether to have children and the roles each partner will take on in the relationship. FSHD may create uncertainty, distress, and tension over whether and when to share information with children about the disease. Disagreements in this area can be upsetting and challenge family relationships.

Depending on when the disease presents and how it progresses, roles and opportunities may change for some or all family members in ways that don't always feel fair. Caring for the needs of the individuals with FSHD may be expensive. Other family members may lose out on social life, education, and career opportunities. FSHD can cause stress, burnout, and isolation for caregivers, who may also feel resentful (and then guilty) about the impact on the family.

Caregiving spouses may avoid speaking up about negative emotions to avoid hurting their FSHD affected partner. The disease may become a source of strife and conflict in the household. Safety concerns for loved ones can clash with the FSHD partner's desire for independence. Balancing protection and independence for a loved one with FSHD is difficult and partners may disagree. Families might have to negotiate these issues repeatedly as the disease progresses.

Studies have found both challenges and resilience in families with a disabled or chronically ill member.[1-2.] Studies also suggest mental health and behavioral challenges for the siblings of disabled children.[3-4] I have so far found no research literature specifically examining the impact of FSHD on spouses or family members.

"Interabled" is a term describing a couple with one disabled and one non-disabled person.5 Jenny and Tony are an interabled couple with a new YouTube channel. Tony has FSHD and uses a wheelchair full-time. They have produced many videos that focus on the challenges of marriage and parenting with disabilities.

Shane Burcaw is a young man with spinal muscular atrophy (SMA) who has written both seriously and humorously about his life challenges.[6] He and his wife, Hannah, have for several years produced a YouTube channel called "Squirmy and Grubs.“ They document their travels together, answer questions, and responding to comments that people post about their relationship. Another writer, Bill Mattlin (also with SMA), wrote a book about love and intimacy within interabled couples. In the book, he interviews several couples (including Shane and Hannah) across the age/relationship duration spectrum about how their differing abilities impact the quality of their marital relationship.[7]

Hard decisions and their consequences

When FSHD shows up in a family, it brings a range of powerful emotional experiences and many hard questions. There's great variety in how the disease makes its presence known within a family. In some couples they identified the disease before the start of the relationship. The couple had some prior knowledge of at least some challenges to be faced in the years ahead. In other families, the diagnosis showed up out of the blue. In about 30% of FSHD cases, the diagnosis shows up with no known family history and is thus a total surprise. When diagnosed early in a relationship, having children may become a topic of discussion. These are often difficult and tinged with powerful feelings. Suppose the disease shows up spontaneously in a child. Sadness, blame, and existential questions may arise for parents in this case. There are no "right" answers to these questions. Honesty, shared values, and love are essential in navigating relationships in families.

Marriages and intimate relationships are difficult to maintain in the best circumstances, as evidenced by the high divorce rate in the US. The everyday pressures and strains of life in our modern world stress relationships. FSHD within the marriage or the family can add to those everyday stresses. FSHD can be expensive, through direct healthcare costs, mobility equipment, modifications to dwellings, and adaptive vehicles. These costs can force hard choices about other things the family may want or need, such as educational opportunities or vacations. FSHD in one spouse may affect employment opportunities and affect family income. Progression of the disease may create other types of tensions as well. The unaffected spouse may worry about the safety of their partner. Partners with FSHD may seek more independence despite the risk of injury. Serious disagreement, conflict, and relational disconnection can result.

Relational tension is a natural part of living life in any family or community. Conflict arises because we each have different perspectives and unique experiences. It's perplexing to understand another person's perspective when ours differs. Yes We all carry our own personal history with us everywhere we go. It's like a backpack that we can't take off - only carry. We are each very aware of why our own particular backpack feels the way it does. Yet, we're not always aware of the backpacks other people carry, and how heavy their load might be. This next exercise is a mindfulness and a perspective-taking exercise designed for couples or family members. This exercise for FSHD families is a variation of one from Dr. Avigail Lev and Dr. Matthew McKay's book *Acceptance and Commitment Therapy for Couples*[8].

EXERCISE 3.7.1 Seeing each other

The following is an exercise done together with a partner. It may expand your awareness of the other person's experience and help build compassion for that experience. Your partner can be your spouse or even an adolescent child. This is a mindfulness exercise. It focuses on perspective taking and seeing and imagining the experience of the person sitting across from you.

It can be challenging, if not impossible, to do an exercise with two people - asking them to at one point have their eyes closed and at other times to gaze into the eyes of the other person —while still trying to read a script out loud. I would suggest three strategies for doing that. First, you can audio-record the entire exercise using the voice recording feature on a smartphone. Second, you can take turns having one individual read a portion and then follow that instruction together. You can alternate the reader back-and-forth. You could also recruit a third person to assist you by reading the italicized sections out loud.

Position yourselves on chairs facing one another, with your knees nearly touching. Sit upright, and lean forward a bit, so that your faces are just a couple feet apart. Take your time with the exercise - don't rush through it even if it feels awkward. Read each section slowly.

1) *As we begin, I would invite each of you to close your eyes and focus your attention on your breathing. Notice how your chest rises and falls as you breathe in and out. Pay attention to the air flowing into and out of your nostrils as you breathe. Notice where in your body you feel your breath most easily? If your attention gets captured by a random thought, notice that without judging yourself, and bring your attention back to your breathing. Keep noticing the experience of breathing. Sit apart from one another with your eyes closed and continue breathing and noticing for the next two minutes*. [Pause 2 minutes]

2) *Now gently open your eyes and raise your gaze to look into your partner's eyes. As you focus on their eyes, try to imagine him/her as a little child. Imagine what he/she might have been like. How did he/she look?* [Pause] *Imagine him/her interacting with his/her family.* [Pause] *Picture the similar phases in life that you each have faced. Picture your partner going to school, getting through puberty, their romantic heartaches. Imagine them becoming an adult, and moving away from home. Notice any emotions, thoughts, or discomfort that might arise, and any urges to laugh, make a joke, or look away. Allow those experiences to be there and continue to focus your gaze on your partner.*

3) *Now close your eyes and return your to attention to breathing, noticing your breath. Pay attention to any thoughts, emotions, or memories coming into your awareness. When you notice them, turn your attention back to your breathing. Breathe in and out for another 30 seconds.* [Pause 30 seconds]

4) *Now re-open your eyes and again look into your partner's eyes. Notice that your partner has had many of their own separate experiences, thoughts, memories, and feelings. He/she has experienced many changing emotions. They've had many feelings, including love, loss, sadness, rejection, hope, loneliness, excitement, and fear. Notice that the person looking into your eyes is a unique person with his/her own experiences and stories. Like you, they are a human being, trying their best. Again, notice what shows up within you as you continue to focus your gaze on your partner.*

5) *Now, again, close your eyes and focus attention on your breathing. Notice the thoughts, emotions, or memories that appear in your awareness. When you notice them, release them gently, and turn your attention back again to your breathing. Gently breathe in and out for another 30 seconds.* [Pause 30 seconds]

6) *Now re-open your eyes and again look into your partner's eyes. This time, I invite you to bring to mind an early memory of first meeting your partner.* [Pause] *What was he/she like when you first met? What were your first experiences together? [Pause] Now bring to mind a time when you saw your partner at his/her best—and when you felt powerful love and pride in your partner.* [Pause] *Now bring to mind a memory of when your partner seemed to be at his/her worst.* [Pause] *Now, this is important! Notice that your partner is still the same person. Sometimes you see him/her at his/her best, sometimes at his/her worst, but he/she remains the same. Sometimes you feel more connected, other times you feel more distant, yet he/she is still the person in front of you with all these distinct qualities. As before, take a moment to notice what shows up inside you: thoughts, emotions, and memories.*

7) *Now, one more time, close your eyes and focus attention on your breathing. Notice any thoughts, emotions, or memories that come into your awareness. When you notice them, release them and gently return to your breathing. Continue breathing in and out for another 30 seconds.* [Pause 30 seconds]

8) *Now, one last time, please open your eyes and gaze into your partner's eyes. Imagine what your partner will look like as he/she gets older. Gaze into his/her eyes and try to picture this face as it ages into the next stage of life and beyond that. Imagine what this will be like for him/her—the experience of his/her body growing older and changing. Yet, also notice that your partner will still be the same human being. Your partner's body will have changed. He/she will have experienced many new things—thoughts, sensations, and emotions. However, they will still be the same human who was there as a child, was there when you met them, was there for the good and the bad, and who is here now. Take a moment to appreciate everything that your partner has experienced and the*

> *person they are. Without speaking, using just your eyes, try to express compassion to them. [Pause] Now bring your attention back to the present moment, and when you are ready, shift your attention back to the room where you are sitting together. You might take a moment to thank your partner for sharing this experience with you.*

Now take some time to talk about that experience together. What did you notice? What sensations, emotions, or thoughts showed up for you? Was the experience pleasant or unpleasant? Did it feel intimate? Is looking at your partner - face to face for an extended period something that you do? How often do you take time to see your partner as a person who carries their own unique history and who struggles just as you do? Would it help to do this more often? Would it help to express compassion and care for their burdens more often?

People have varied experiences when doing this exercise with a partner. Some feel a deep sense of connection when they do that exercise. For others, the experience can feel disconcerting or even uncomfortable. What are the important values that you have that affect how are you live in relationship with your partner, and with other family members? What actions can you take today that might express those values?

* * *

Changed family roles and responsibilities

Two and a half millennia ago, the Greek philosopher Heraclitus said, "*Change is the only constant in life*." In our modern age, the pace of change has increased to where it often feels difficult to keep up. Change in physical functioning is perhaps the hallmark symptom of a progressive disease such as FSHD. But the change that FSHD brings not only affects the individual with the disease but also every member of the family. As the disease steals away physical abilities from the affected family member, the roles, responsibilities, and specific activities of other family members often change as well.

For example, as FSHD impairs a person's ability to reach above their head, they may no longer be able to put away dishes from the dishwasher into upper cabinets. Perhaps another family member will need to take on that chore. Maybe someone in the family will have to re-organize the kitchen cabinets to make them more accessible. New roles and responsibilities may cause conflict and emotions. When one person's physical limitations impact and force changes in another person's activities, it can feel unfair. But, because those changes came because of a disease that no one has control over - family members might not express those feelings, only feel them. Carrying around unexpressed feelings of resentment or frustration can lead to conflicts that are not about what they seem to be about. Families dealing with FSHD need communication and perspective-taking skills.

Humans are at their core social creatures. We need relationships with and support from other humans. Our species survived on this planet through cooperation and shared struggle. Those of us who are fortunate find much of that love and support within our immediate family. Others may have to search to find those connections with friends and those who share common values and interests. Those with FSHD need to build strong relationships with family and friends. It is also essential for those who provide caregiving for people with FSHD

Developmental challenges throughout the lifespan

It's well known that people have different life experiences and challenges at different periods of their lives. It's beyond this book's scope to address all these challenges, and there are other books on ACT directed toward them. But I want to mention two specific time frames that deserve attention related to the impact of FSHD.

The Turmoil of Adolescence

We often associate adolescence with some of the highest and lowest times of a person's life. It's a time when one experiences many "firsts," such as first love, first job, first car and so on. It's a time when the need and desire for autonomy and independence may create tension with parents. It's also a time when hormonal changes create emotional volatility and behavioral impulsivity that can be painful. Sometimes, adolescent experiences may cause enduring and even lifelong consequences. Adolescents with FSHD experience all the above besides their disease issues. Adolescents with FSHD symptoms can lose confidence, withdraw from social activities, and feel depressed or angry.

The adolescent may angrily call into question earlier parental decisions about when and how to test for FSHD. I've spoken with parents of FSHD adolescents whose children blame them for being born. They stop communicating with parents and sometimes struggle with suicidal thoughts. Adolescents with visibly progressing FSHD symptoms may feel emotional pain, fear, and disconnection. In these instances, parents should try to secure professional mental health support and to find locate FSHD peers for support. It may also help to find slightly older mentors within the FSHD community who can help guide adolescents through this challenging time.

Louise Hayes and Joseph Ciarrochi are psychologists with expertise in both ACT and the challenges of adolescence. They have co-authored two beneficial books designed to teach ACT principles to adolescents[9-10]. Though these books don't focus on FSHD or on disability, teens may still find them useful in navigating through these tough challenges. Here is a link to their most recent book for teens.

Aging with FSHD

For most of us, aging involves coming to terms with the prospect of physical decline, loss of loved ones, and death. Having to face these things can feel frightening and lonely for each of us. Disease processes and subsequent disability from medical problems often complicate normal aging. Diseases like diabetes, obesity, sleep apnea, high blood pressure, heart disease, and cancer can diminish quality of life. Health limitations created by these problems can worsen disability and add stress. Because FSHD is a progressive disease, its most powerful impact will always occur during one's later years. Other co-occurring diseases can worsen that impact.

Developmental challenges impact the whole family, not just the individual. Existing challenges can worsen and additional problems can arise because of FSHD..

Aging provides not only hard challenges, it also brings new opportunities. For me, aging and retirement have freed me to reevaluate how I spend my time and to engage in the things that matter to me - my values. Providing leadership, and being involved in the local FSHD Society chapter, is one avenue my wife and I are living and engaging in our shared values. We get to love and care for our kids and grandkids in retirement. COVID-19 restrictions and distance have made some opportunities only virtual during some of this time.

Engaging with the wider FSHD community

In this book, I've said that people with FSHD and their families often feel isolated. FSHD families don't have to endure this experience alone. I can't tell you enough how meaningful involvement in the broader community of FSHD has been for me and my wife. The FSHD Society has a growing chapter program throughout the country. There are currently 27 local chapters scattered throughout the US. Even before COVID-19, many of these local chapters hosted regular meetings that included the option to take part remotely. My wife and I have met many new friends and received much social support, encouragement, and helpful advice from other FSHD society members. To find a local chapter in your area and to learn more about the chapter program, click here.

The last exercise in this book is to identify important values for being part of a community with FSHD. I adapted the following activity for FSHD from one created by Jenna T. LeJeune and. Jason Luoma.

EXERCISE 3.7.2 Your FSHD community guidelines

Instructions: Most societies and cultures choose a set of guiding principles they feel their members should live by. Judeo Christian culture laid out the 10 Commandments thousands of years ago. Two centuries ago, the Declaration of Independence and the Bill of Rights set forward guiding principles for U.S. society. Most organizations set rules and guidelines. In today's society, each social media platform has different posting guidelines.. These sorts of guidelines can be beneficial because they can help us get along in a communal society. Often in communities and on social media, the emphasis is on punishing misconduct rather **tha**n advocating positive actions in communities and social **med**ia..

ACT focuses on moving towards our chosen direction to create the life we want.

For this exercise, I invite you to write your personal set of community guidelines for living within your extended FSHD family. I invite you to make this a family exercise. Sit down together, Talk about what’s important in your family. Make sure you hear from everyone. These are not to be statements about how not to live. Instead, they should be declarations of what you would choose to make important in your life together, what you desire your family life to be about. So instead of writing these guidelines starting with "Thou shall not…” remember, to engage, you are free to choose. You could choose to stand for anything, “Love” “Justice” “Equality” “Adventure.” Your family gets to be the designer of your own community.

Take a few moments to consider the values you would choose to live by to enhance relationships with friends, family members, neighbors, work colleagues, and even strangers.

- As part of my FSHD community we choose to stand for:

  ____

- As part of my FSHD community we choose to stand for:

____

- As part of my FSHD community we choose to stand for:
  ____
- As part of my FSHD community we choose to stand for:
  ____
- As part of my FSHD community we choose to stand for:
  ____
- As part of my FSHD community we choose to stand for:
  ____
- As part of my FSHD community we choose to stand for:
  ____
- As part of my FSHD community we choose to stand for:
  ____
- As part of my FSHD community we choose to stand for:
  ____
- As part of my FSHD community we choose to stand for: ____________________________

After you have finished, take a few moments, read your statements carefully and reflect on the following questions. *As you look these over, would other people reading your guidelines clearly understand what your family stands for? What's most important to you? Would they understand the people you are by reading these statements? Would you be proud to be a part of a community that stood for these values?*

Now try to identify some specific actions that you each could take today to stand for those values. Write those actions down and commit to doing them today.

Conclusion

Coming to terms with FSHD will always be an unfinished process because the disease keeps progressing throughout our lives. As our disease progresses, we must continue to adapt. To live fully, to make the most of all the time we have, we must focus our present moments on doing the things that matter most to us. This is the path to vital living. If you've read this far in the book, I hope that you've received some benefit that will help you on your journey.

In Part One of the book, I summarized the challenges of doing research with a rare disease such as FSHD. I've tried to share what I learned from research about how FSHD affects mental health. FSHD research funding has almost exclusively targeted biological disease mechanisms and treating physical symptoms. Because of this, research on mental health in FSHD is sparse. I've included research on the mental health impact of other chronic progressive diseases. I hope my review provided at least an introduction to this research.

In Part Two, I summarized Acceptance and Commitment Therapy (ACT). This is a well researched, evidence-based approach to psychotherapy. ACT is trans-diagnostic. By this I mean, ACT doesn't focus on eliminating the symptoms of particular psychological disorders. Rather, ACT aims to increase psychological flexibility for anyone, not just those with mental health issues. Steve Hayes, one of the primary creators of ACT, describes psychological flexibility this way in his most recent book.[1]

> *"Psychological flexibility is the ability to feel and think with openness, to attend voluntarily to your experience of the present moment, and to move your life in directions that are important to you, building habits that allow you to live in ways reflecting your values and aspirations. It's about learning not to turn away from what is painful, instead turning toward your suffering in order to live a life full of meaning and purpose."*

Over 600 randomized clinical trials (RCT) establish the scientific basis of ACT. These range across a very wide range of mental health problems and life difficulties. ACT provides a firm base to support each of us struggling with FSHD. It can also give support for family members and caregivers who each have their own unique struggles.

Part Three of this book focuses on applying ACT principles to common challenges faced by FSHD patients and their families. There are seven modules included in Part Three. Not every module will apply to everyone, but hopefully you have found things to assist you on your personal journey. I would invite you to come back to the book from time to time as your disease changes and life brings new challenges. Many of the exercises in one module, you can apply equally well to other challenges. You may benefit from looking at all the exercises, even in modules that don't seem to apply directly to your current needs. Taking your time in this process, and returning to the book again and again, may be a strategy for getting the most benefit.

I know that there is a stigma in our country about seeking mental health support. I also know that psychotherapy is expensive, and not always well-covered by health insurance plans. Co-pays can be high, and the number of sessions limited. I also know that financial resources for people with disabilities are often limited and focused on more pressing needs. There are really two reasons I chose to self-publish this book. Publishers felt the FSHD community was too small for financial viability, as I wanted to focus just on FSHD. I also wanted freedom to make the book available to every person with FSHD who might be interested in and benefit from it.

There is a wealth of additional information available about ACT. There is a professional organization called the Association for Contextual Behavioral Science (ACBS). This organization hosts professional conferences and educational events to train ACT therapists and researchers. The organization has a professional journal where they publish ACT-related research. On the Home page of the ACBS website, there is a 'Find an ACT therapist' button - that will allow you to search for a provider of ACT in your area. There are other ways to find an ACT therapist. The Psychology Today online web portal also has a find a therapist box on the Home page. You can put in your city or ZIP Code and press search. This will bring up a general list of therapists - with several drop-down buttons above the pictures. Click on the Types of Therapy button, and you will see Acceptance and Commitment Therapy (ACT) listed as one of the therapeutic modalities. Clicking on that link will bring up a list of the local therapists who provide ACT.

There are also many self-help books on ACT for many life problems. You can search online bookstores such as Amazon or Barnes & Noble using "Acceptance and Commitment Therapy" as a search term to find these books. You can also find published research on ACT using Google Scholar.

Throughout my life, avoidance has been a frequent but unhelpful way of coping with my FSHD. This led to feeling alone and isolated with my disease. Trying to cope with the progression of this terrible disease without supportive friends and helpful resources is hard. In 2016, when I finally met other people with FSHD for the first time, my sense of hopefulness grew. That came about because of the work of the FSHD Society. They sponsored the event where that happened. Since that time, I've chosen to volunteer as co-director of the San Francisco Bay Area chapter of the FSHD Society. It has helped me feel a part of a powerful community effort to find a cure for this disease. It has given me hope to continue forward in my struggle with this disease. I would encourage anyone reading this book to take the simple step of connecting with others who share your struggle. You are not and do not have to be alone on this journey!

References

Introduction

1. White, L. *The graceful art of falling*. Bloomington, IN: AuthorHouse. 2013.

Part One

1. Statland J, Tawil R. Facioscapulohumeral muscular dystrophy. Neurol Clin 2014; 32: 721-8, ix.
2. Deenen JCW, Arnts H, Van Der Maarel SM, et al. Population-based incidence and prevalence of facioscapulohumeral dystrophy. *Neurology*. 2014;83(12):1056-1059. doi:10.1212/WNL.0000000000000797.
3. Institute of Medicine. *Rare Diseases and Orphan Products: Accelerating Research and Development*. (Field MJ, Boat TF, eds.). Institute of Medicine of the National Academies; 2011:1-442.
4. Johnson NE, Quinn C, Eastwood E, Tawil R, Heatwole CR. Patient-identified disease burden in facioscapulohumeral muscular dystrophy. *Muscle Nerve*. 2012;46(6):948-950. doi:10.1002/mus.23529.
5. Hamel J, Johnson N, Tawil R, et al. Patient-Reported Symptoms in Facioscapulohumeral Muscular Dystrophy (PRISM-FSHD). *Neurology*. 2019;93(12):e1180-e1192. doi:10.1212/WNL.0000000000008123.
6. National Center for Health Statistics. *Health, United States, 2006 with chartbook on trends in the health of Americans*. Hyattsville, MD: NCHS. 2006.
7. Gerrits, M, van Oppen, P, van Marwijk, H, Penninx, B, van der Horst, H. Pain and the onset of depressive and anxiety disorders. *Pain*. 2014;155(1). https://doi.org/10.1016/j.pain.2013.09.005
8. Alschuler KN, Jensen MP, Goetz MC, Smith AE, Verrall AM, Molton IR. Effects of pain and fatigue on physical functioning and depression in persons with muscular dystrophy. *Disability and Health Journal*. 2012;5(4):277-283. doi:10.1016/j.dhjo.2012.07.002.
9. Graham CD, Rose MR, Grunfeld EA, Kyle SD, Weinman J. A systematic review of quality of life in adults with muscle disease. *J Neurol*. 2011;258(9):1581-1592. doi:10.1007/s00415-011-6062-5.
10. Padua L, Aprile I, Frusciante R, et al. Quality of life and pain in patients with facioscapulohumeral muscular dystrophy. *Muscle Nerve*. 2009;40(2):200-205. doi:10.1002/mus.21308.
11. Morís G, Wood L, FernáNdez-Torrón R, et al. Chronic pain has a strong impact on

quality of life in facioscapulohumeral muscular dystrophy. *Muscle Nerve*. 2017;75:550. doi:10.1002/mus.25991.

12. Kalkman JS, Schillings ML, van der Werf SP, et al. Experienced fatigue in facioscapulohumeral dystrophy, myotonic dystrophy, and HMSN-I. *Journal of Neurology, Neurosurgery & Psychiatry*. 2005;76(10):1406-1409. doi:10.1136/jnnp.2004.050005.
13. Kalkman JS, Schillings ML, Zwarts MJ, van Engelen BGM, Bleijenberg G. The development of a model of fatigue in neuromuscular disorders: a longitudinal study. *Journal of Psychosomatic Research*. 2007;62(5):571-579. doi:10.1016/j.jpsychores.2006.11.014.
14. Voet N, Bleijenberg G, Hendriks J, et al. Both aerobic exercise and cognitive-behavioral therapy reduce chronic fatigue in FSHD: An RCT. *Neurology*. 2014;83(21):1914-1922. doi:10.1212/WNL.0000000000001008.
15. Kalkman JS, Schillings ML, Zwarts MJ, van Engelen BGM, Bleijenberg G. Psychiatric disorders appear equally in patients with myotonic dystrophy, facioscapulohumeral dystrophy, and hereditary motor and sensory neuropathy type I. *Acta Neurol Scand*. 2007;115(4):265-270. doi:10.1111/j.1600-0404.2006.00737.x.
16. Bungener C, Jouvent R, Delaporte C. Psychopathological and emotional deficits in myotonic dystrophy. *Journal of Neurology, Neurosurgery & Psychiatry*. 1998;65(3):353-356.
17. Meade O, Buchanan H, Coulson N. The use of an online support group for neuromuscular disorders: a thematic analysis of message postings. *Disabil Rehabil*. 2018;40(19):2300-2310. doi:10.1080/09638288.2017.1334239.
18. Petrie KJ, Weinman J. Patients' Perceptions of Their Illness. *Current Directions in Psychological Science*. 2012;21(1):60-65. doi:10.1177/0963721411429456.
19. Chilcot J, Wellsted D, Farrington K. Illness perceptions predict survival in haemodialysis patients. Am J Nephrol 2011; 33: 358-363.
20. Graham CD, Rose MR, Hankins M, Chalder T, Weinman J. Separating emotions from consequences in muscle disease: Comparing beneficial and unhelpful illness schemata to inform intervention development. *Journal of Psychosomatic Research*. 2013;74(4):320-326. doi:10.1016/j.jpsychores.2012.09.012.
21. Graham CD, Weinman J, Sadjadi R, et al. A multicentre postal survey investigating the contribution of illness perceptions, coping and optimism to quality of life and mood in adults with muscle disease. *Clinical Rehabilitation*. 2014;28(5):508-519. doi:10.1177/0269215513511340.
22. Graham CD, Gouick J, Ferreira N, Gillanders D. The Influence of Psychological Flexibility on Life Satisfaction and Mood in Muscle Disorders. *Rehabilitation Psychology*. 2016;61(2):210-217. doi:10.1037/rep0000092.supp.
23. Graham CD, Rose MR, Grunfeld EA, Kyle SD, Weinman J. A systematic review of quality of life in adults with muscle disease. *J Neurol*. 2011;258(9):1581-1592. doi:10.1007/

s00415-011-6062-5.

24. Graham CD, Gouick J, Krahé C, Gillanders D. A systematic review of the use of Acceptance and Commitment Therapy (ACT) in chronic disease and long-term conditions. *Clinical Psychology Review*. 2016;46(C):46-58. doi:10.1016/j.cpr.2016.04.009.
25. Graham CD, O'Hara DJ, Kemp S. A case series of Acceptance and Commitment Therapy (ACT) for reducing symptom interference in functional neurological disorders. *Clin Psychol Psychother*. 2018;25(3):489-496. doi:10.1002/cpp.2174.
26. Graham CD, Chalder T, Rose MR, Gavriloff D, McCracken LM, Weinman J. A pilot case series of a brief acceptance and commitment therapy (ACT)-based guided self-help intervention for improving quality of life and mood in muscle disorders. *tCBT*. 2017;10:167–26. doi:10.1017/S1754470X17000022.
27. Rose MR, Norton S, Vari C, et al. Acceptance and Commitment Therapy for Muscle Disease (ACTMus): protocol for a two-arm randomised controlled trial of a brief guided self-help ACT programme for improving quality of life in people with muscle diseases. *BMJ Open*. 2018; 8(10):e022083–10. doi:10.1136/bmjopen-2018-022083.
28. Rose, M., Graham, Christopher D., O'Connell, Nicola, Vari, Chiara, Edwards, Victoria, Taylor, Emma, McCracken, Lance M., Radunovic, Aleksander, Chalder, Trudie. A randomised controlled trial of Acceptance and Commitment Therapy for improving quality of life in people with muscle diseases. *Psychological Medicine*. 2022; 1–14. doi: 10.1017/S0033291722000083

Part Two

1. Kessler RC, Berglund P, Demler O, Jin R, Merikangas KR, Walters EE. Lifetime prevalence and age-of-onset distributions of DSM-IV disorders in the National Comorbidity Survey Replication. *Arch Gen Psychiatry*. 2005;62(6):593-602. doi:10.1001/archpsyc.62.6.593.
2. Hayes, SC., Strosahl, KD, Kirk, D., & Wilson, KG. *Acceptance and Commitment Therapy*, Second Edition (p. 4). Kindle Edition. New York, NY. Guilford Publications. 2012.
3. Westrup, D. *Advanced Acceptance and Commitment Therapy: The Experienced Practitioner's Guide to Optimizing Delivery* (p.9). Kindle Edition. Oakland , CA. New Harbinger Publications, Inc. 2014.
4. Kabat-Zinn, J. *Wherever you go, there you are: Mindfulness meditation in everyday life.* New York: Hyperion Books. 1994.
5. Hayes, SC., Strosahl, KD, Kirk, D., & Wilson, KG. *Acceptance and Commitment Therapy*, Second Edition (p. 272). Kindle Edition. New York, NY. Guilford Publications. 2012.
6. Robb, H. Opening up: Acceptance and defusion. In: Nieuwsma JA, Walser RD, Hayes SC, eds. *ACT for Clergy and Pastoral Counselors*. 2016:185-202.
7. Hayes, SC., Strosahl, KD, Kirk, D., & Wilson, KG. *Acceptance and Commitment Therapy*,

Second Edition (p. 246). Kindle Edition. New York, NY. Guilford Publications. 2012.

8. Blackledge JT. Disrupting verbal processes: Cognitive defusion in acceptance and commitment therapy and other mindfulness-based psychotherapies. *Psychological Record*. 2007;57(4):555-577.
9. Titchener, E. B. *A text-book of psychology.* New York, NY. MacMillan. 1915.
10. Hayes, SC., Strosahl, KD, & Wilson, K. *Acceptance and commitment therapy*: An experimental approach to behavior change. *New York, NY. Guilford Press. 1999.*
11. Kabat-Zinn, J. *Full catastrophe living: Using the wisdom of your body and mind to face stress, pain and illness*. New York, NY: Delacorte. 1990.
12. Walser, RD, Westrup, D. *Acceptance and commitment therapy for the treatment of post-traumatic stress disorder and trauma-related problems: A practitioner's guide to using mindfulness and acceptance strategies*. Oakland, CA: New Harbinger Publications, Inc. 2007.

Part Three

1. Graham CD, Rose MR, Hankins M, Chalder T, Weinman J. Separating emotions from consequences in muscle disease: Comparing beneficial and unhelpful illness schemata to inform intervention development. *Journal of Psychosomatic Research*. 2013;74(4):320-326. doi:10.1016/j.jpsychores.2012.09.012.
2. Graham CD, Weinman J, Sadjadi R, et al. A multicentre postal survey investigating the contribution of illness perceptions, coping and optimism to quality of life and mood in adults with muscle disease. *Clinical Rehabilitation*. 2014;28(5):508-519. doi:10.1177/0269215513511340.
3. Lorig KR, Sobel, DS, Ritter PL, Laurent, D, Hobbs, M. Effect of a self-management program on patients with chronic disease. *Effective Clinical Practice*, 4, 2001,pp. 256-262.
4. Podsiadlo D, Richardson S. The Timed Up & Go: A test of basic functional mobility for frail elderly persons. *J Am Geriatr Soc.* 1991;39:142-148.
5. Enright PL, McBurnie MA, Bittner V, Tracy RP, McNamara R, Arnold A, et al. The 6 minute walk test: a quick measure of functional status in elderly adults. Chest 2003;123(2):387–398
6. Kroenke, K, Spitzer, RL, Williams, JB. The phq-9. *Journal of general internal medicine*, 2001;16(9):606-613.
7. Bond FW, Hayes SC, Baer RA, et al. Preliminary psychometric properties of the Acceptance and Action Questionnaire-II: a revised measure of psychological inflexibility and experiential avoidance. *Behavior Therapy*. 2011;42(4):676-688. doi:10.1016/j.beth.2011.03.007.
8. Francis AW, Dawson DL, Golijani-Moghaddam N. The development and validation of the Comprehensive assessment of Acceptance and Commitment Therapy processes

(CompACT). *Journal of Contextual Behavioral Science*. 2016;5:134-145. doi:10.1016/ j.jcbs.2016.05.003.

Module 1

1. Walser, RD, Westrup, D. *Acceptance and commitment therapy for the treatment of post-traumatic stress disorder and trauma-related problems: A practitioner's guide to using mindfulness and acceptance strategies*. Oakland, CA: New Harbinger Publications, Inc. 2007.
2. Hayes, SC. Strosahl, KD, & Wilson, K. G. *Acceptance and Commitment Therapy: An Experiential Approach to Behavior Change*. New York: Guilford Press. 1999.
3. LeJeune, J, Luoma, JB. *Values in Therapy: A clinician's guide to helping clients explore values, increase psychological flexibility and live a more meaningful life*. Oakland: Context Press, New Harbinger Publications. 2019.

Module 2

1. Worry. Merriam-Webster.com. 2020. https://www.merriam-webster.com Accessed June 13, 2020.
2. LeJeune, C. The Worry Trap (p. 30). Oakland: New Harbinger Publications. Kindle Edition. 2007.
3. Hayes, SC., Strosahl, KD, Kirk, D., & Wilson, KG. *Acceptance and Commitment Therapy*, Second Edition (p. 250). Kindle Edition. New York, NY. Guilford Publications. 2012.

Module 3

1. Hamel J, Johnson N, Tawil R, et al. Patient-Reported Symptoms in Facioscapulohumeral Muscular Dystrophy (PRISM-FSHD). *Neurology*. 2019;93(12):e1180-e1192. doi:10.1212/WNL.0000000000008123.
2. Tolkien, J. R. R. The Hobbit, Or, There and Back Again. 1st Revised Edition. New York: Ballantine Books. 1966.
3. Walser, RD, Westrup, D. *Acceptance and commitment therapy for the treatment of post-traumatic stress disorder and trauma-related problems: A practitioner's guide to using mindfulness and acceptance strategies*. Oakland, CA: New Harbinger Publications, Inc. 2007.

Module 4

1. Kalkman JS, Schillings ML, Zwarts MJ, van Engelen BGM, Bleijenberg G. The development of a model of fatigue in neuromuscular disorders: a longitudinal study. *Journal of Psychosomatic Research*. 2007;62(5):571-579. doi:10.1016/ j.jpsychores.2006.11.014.
2. Voet N, Bleijenberg G, Hendriks J, et al. Both aerobic exercise and cognitive-behavioral therapy reduce chronic fatigue in FSHD: An RCT. *Neurology*. 2014;83(21):1914-1922. doi:10.1212/WNL.0000000000001008.
3. Westrup, D. *Advanced Acceptance and Commitment Therapy: The Experienced Practitioner's Guide to Optimizing Delivery* (p.9). Kindle Edition. Oakland , CA. New

Harbinger Publications, Inc. 2014.

4. Stoddard, JA, Afari, N. *The Big Book of ACT Metaphors: A Practitioner's Guide to Experiential Exercises and Metaphors in Acceptance and Commitment Therapy* . Oakland: New Harbinger Publications. 2014.

Module 5

1. Vadivelu, N., Kai, A.M., Kodumudi, G., Babayan, K., Fontes, M., & Burg, M. M. (2017). Pain and Psychology—A Reciprocal Relationship. Ochsner Journal, 17, 173-180.
2. Hamel J, Johnson N, Tawil R, et al. Patient-Reported Symptoms in Facioscapulohumeral Muscular Dystrophy (PRISM-FSHD). *Neurology*. 2019;93(12):e1180-e1192. doi:10.1212/WNL.0000000000008123.
3. Morís G, Wood L, FernáNdez-Torrón R, et al. Chronic pain has a strong impact on quality of life in facioscapulohumeral muscular dystrophy. *Muscle Nerve*. 2017;75:550. doi:10.1002/mus.25991.
4. Bushby KM, Pollitt C, Johnson MA, Rogers MT, Chinnery PF. Muscle pain as a prominent feature of facioscapulohumeral muscular dystrophy (FSHD): four illustrative case reports. *Neuromuscular Disorders*. 1998;8(8):574-579. doi:10.1016/s0960-8966(98)00088-1.
5. Rosenblum A, Marsch LA, Joseph H, Portenoy RK. Opioids and the treatment of chronic pain: controversies, current status, and future directions. *Exp Clin Psychopharmacol*. 2008;16(5):405-416. doi:10.1037/a0013628
6. McCracken, L. ACT for Chronic pain. *Retreived from: https://www.div12.org/wp-content/uploads/2015/06/ACT-for-Chronic-Pain-manual-McCracken.pdf*. 2012, 46pp.
7. Hayes, SC., Strosahl, KD, Kirk, D., & Wilson, KG. *Acceptance and Commitment Therapy*, Second Edition (p. 276). Kindle Edition. New York, NY. Guilford Publications. 2012.
8. Hayes, SC. A Liberated Mind. London: Penguin Publishing Group. 2019.
9. Stoddard, JA, Afari, N. The Big Book of ACT Metaphors: A Practitioner's Guide to Experiential Exercises and Metaphors in Acceptance and Commitment Therapy . Oakland: New Harbinger Publications. 2014.

Module 6

1. Hales, CM, Carroll, MD, Fryar, CD, and Ogden, CL. Prevalence of Obesity and Severe Obesity Among Adults: United States, 2017–2018. CDC: NCHS Data Brief No. 360, February 2020. https://www.cdc.gov/nchs/products/databriefs/db360.htm. Accessed May 24, 2020.

2. Cruz-Jentoft AJ, Baeyens JP, Bauer JM, et al. Sarcopenia: European consensus on definition and diagnosis: Report of the European Working Group on Sarcopenia in Older People. *Age and Ageing*. 2010;39(4):412-423. doi:10.1093/ageing/afq034.

3. Baumgartner RN. Body composition in healthy aging. *Ann N Y Acad Sci*. 2000;904(1):437-448. doi:10.1111/j.1749-6632.2000.tb06498.x.

4. Vera K, McConville M, Kyba M, Keller-Ross M. Sarcopenic Obesity in Facioscapulohumeral *Muscular Dystrophy*. Preprint at https://www.researchsquare.com/article/rs-14397/v1. (February 2020).

5. Lillis J, Luoma JB, Levin ME, Hayes SC. Measuring Weight Self-stigma: The Weight Self-stigma Questionnaire. *Obesity* 2010; 18: 971-976.

6. Lillis J, Hayes SC, Bunting K, Masuda A. Teaching Acceptance and Mindfulness to Improve the Lives of the Obese: A Preliminary Test of a Theoretical Model. *Annals of Behavioral Medicine* 2009; 37: 58-69.

7. Stoddard, JA, Afari, N. *The Big Book of ACT Metaphors: A Practitioner's Guide to Experiential Exercises and Metaphors in Acceptance and Commitment Therapy* . Oakland: New Harbinger Publications. 2014.

ACT for caregivers and family members

1. Holroyd J, Guthrie D. Family stress with chronic childhood illness: cystic fibrosis, neuromuscular disease, and renal disease. *J Clin Psychol*. 1986;42(4):552-561. doi:10.1002/1097-4679(198607)42:4<552::aid-jclp2270420403>3.0.co;2-8.
2. Chen J-Y, Clark M-J. Family function in families of children with Duchenne muscular dystrophy. *Fam Community Health*. 2007;30(4):296-304. doi:10.1097/01.FCH.0000290542.10458.f8.
3. Sharpe D, Rossiter L. Siblings of children with a chronic illness: a meta-analysis. *J Pediatr Psychol*. 2002;27(8):699-710. doi:10.1093/jpepsy/27.8.699.
4. Read J, Kinali M, Muntoni F, Weaver T, Garralda ME. Siblings of young people with Duchenne muscular dystrophy--a qualitative study of impact and coping. *Eur J Paediatr Neurol*. 2011;15(1):21-28. doi:10.1016/j.ejpn.2010.07.006.
5. Mattlin, B. Help! I think I created a word! April 8, 2019. Available at: https://facingdisability.com/blog/help-i-think-i-created-a-word. Accessed on September 15, 2020.
6. Burcaw, S. Laughing at My Nightmare. New York: Roaring Brook Press. 2014.
7. Mattlin, B. In Sickness and in Health: Love, Disability, and a Quest to Understand the Perils and Pleasures of Interabled Romance. Boston: Beacon Press. 2018
8. Lev, A, & McKay, M. Acceptance and Commitment Therapy for Couples. Oakland, CA: New Harbinger Publications; 2017.
9. Ciarrochi, JV, Hayes,LL, and Bailey, A. Get Out of Your Mind and Into Your Life for Teens: A Guide to Living an Extraordinary Life. Oakland, CA: New Harbinger; 2012.
10. Hayes, LL, Ciarrochi, JV. The Thriving Adolescent: Using Acceptance and Commitment Therapy and Positive Psychology to Help Teens Manage Emotions, Achieve Goals, and Build Connection. Oakland, CA: Context Press; 2015.

Conclusion

1. Hayes, SC. A Liberated Mind. London: Penguin Publishing Group. 2019.

Appendices

1. Bond FW, Hayes SC, Baer RA, et al. Preliminary psychometric properties of the Acceptance and Action Questionnaire-II: a revised measure of psychological inflexibility and experiential avoidance. *Behavior Therapy*. 2011;42(4):676-688. doi:10.1016/j.beth.2011.03.007.

2. Francis AW, Dawson DL, Golijani-Moghaddam N. The development and validation of the Comprehensive assessment of Acceptance and Commitment Therapy processes (CompACT). *Journal of Contextual Behavioral Science*. 2016;5:134-145. doi:10.1016/j.jcbs.2016.05.003.

3. Carney CE, Buysse DJ, Ancoli-Israel S, et al. The consensus sleep diary: standardizing prospective sleep self-monitoring. *Sleep*. 2012;35(2):287-302. doi:10.5665/sleep.1642.
4. Lillis J, Luoma JB, Levin ME, Hayes SC. Measuring Weight Self-stigma: The Weight Self-stigma Questionnaire. *Obesity*. 2009;18(5):971-976. doi:10.1038/oby.2009.353.

Photo Sources

Caption Link	Image Source
Cover	Drescher, Susan. (Photographer). (2014, February). *Off Highway 1* [digital image]. Drescher, Kent. (Photographer). (2014, February). *Bridge off Highway 1* [digital image].
Empty Swing	Free-Photos (Photographer). (2020, April) *Swing-846077_1920.jpg* [digital image]. Retrieved from https://pixabay.com/
Feather	Godfrida, Maria (Photographer). (2020, April) *Feather-1078424_1920.jpg* [digital image]. Retrieved from https://pixabay.com/
Cactus	Palla, Martin (Photographer). (2020, April) *Cactus-1397679_1920.jpg* [digital image]. Retrieved from https://pixabay.com/
Forest Stream	272447 (Photographer). (2020, April) *Forest-1163565_1920.jpg* [digital image]. Retrieved from https://pixabay.com/
Glass of Milk	Pezibear (Photographer). (2020, April) *Glass-1379822_1920.jpg* [digital image]. Retrieved from https://pixabay.com/
Classical Guitar	Edwin Valencia (Photographer). (2020, April) *guitar-3870970_1920.jpg* [digital image]. Retrieved from https://pixabay.com/
Teddy Bear	Congerdesign (Photographer) (2020, April) *teddy-562960_1920.jpg* [digital image]. Retrieved from https://pixabay.com/
Cake	Guvo59 (Photographer). (2020, May) *muffin-3607780_1920.jpg* [digital image]. Retrieved from https://pixabay.com/
River Rocks	Sarajuggernaut (Photographer). (2020, June) *stones-167089_1920.jpg* [digital image]. Retrieved from https://pixabay.com/
Rock	Melissa Tomczak (Photographer). (2020, June) *021_FreeRange_04_2012.jpg* [digital image]. Retrieved from https://freerangestock.com/
Tea	Congerdesign (Photographer). (2020, August) *tee-1740871_1920.jpg* [digital image]. Retrieved from https://pixabay.com/
Rope Bridge	Coffee (Photographer). (2020, August) adventure-72127_1280.jpg [digital image].Retrieved from https://pixabay.com/
Key Chain	Torstensimon (Photographer). (2022, January) key-g796453a5f_1920.jpg [digital image].Retrieved from https://pixabay.com/

Photographer, F.M. (Photographer). (Year, Month Date of Publication). *File name of Photograph* [digital image]. Retrieved from URL

Appendices

Appendix 1

Stanford HAQ 8-Item Disability Scale

Please CIRCLE the one best answer for your abilities.

At this moment, are you able to:	**With *NO* difficulty 0**	**With *SOME* difficulty 1**	**With *MUCH* difficulty 2**	***UNABLE* to do 3**
1. Dress yourself, including tying shoelaces and doing buttons?	**0**	**1**	**2**	**3**
2. Get in and out of bed?	**0**	**1**	**2**	**3**
3. Lift a full cup or glass to your mouth?	**0**	**1**	**2**	**3**
4. Walk outdoors on flat ground?	**0**	**1**	**2**	**3**
5. Wash and dry your entire body?	**0**	**1**	**2**	**3**
6. Bend down to pick up clothing from the floor?	**0**	**1**	**2**	**3**
7. Turn faucets on and off?	**0**	**1**	**2**	**3**
8. Get in and out of a car?	**0**	**1**	**2**	**3**

Scoring

Score the number circled for each item. If more than one consecutive number is circled for one item, code the higher number (more difficulty). If responses are not consecutive, code as blank. The disability index is the mean of the eight items. If more than 2 items are blank, do not score the index.

Reference

Lorig KR, Sobel, DS, Ritter PL, Laurent, D, Hobbs, M. Effect of a self-management program on patients with chronic disease. Effective Clinical Practice, 4, 2001,pp. 256-262.

This scale is free to use without permission
Self-Management Resource Center
www.selfmanagementresource.com

Appendix 2

PATIENT HEALTH QUESTIONNAIRE-9 (PHQ-9)

Over the last 2 weeks, how often have you been bothered by any of the following problems? *(Use "✔" to indicate your answer)*	**Not at all**	**Several days**	**More than half the days**	**Nearly every day**
1. Little interest or pleasure in doing things	0	1	2	3
2. Feeling down, depressed, or hopeless	0	1	2	3
3. Trouble falling or staying asleep, or sleeping too much	0	1	2	3
4. Feeling tired or having little energy	0	1	2	3
5. Poor appetite or overeating	0	1	2	3
6. Feeling bad about yourself — or that you are a failure or have let yourself or your family down	0	1	2	3
7. Trouble concentrating on things, such as reading the newspaper or watching television	0	1	2	3
8. Moving or speaking so slowly that other people could have noticed? Or the opposite — being so fidgety or restless that you have been moving around a lot more than usual	0	1	2	3
9. Thoughts that you would be better off dead or of hurting yourself in some way	0	1	2	3

FOR OFFICE CODING _0_ + _____ + _____ + _____

=Total Score: _____

If you checked off any problems, how difficult have these problems made it for you to do your work, take care of things at home, or get along with other people?

Not difficult at all	**Somewhat difficult**	**Very difficult**	**Extremely difficult**
☐	☐	☐	☐

Developed by Drs. Robert L. Spitzer, Janet B.W. Williams, Kurt Kroenke and colleagues, with an educational grant from Pfizer Inc. No permission required to reproduce, translate, display or distribute.

Appendix 3

AAQ-II

Below you will find a list of statements. Please rate how true each statement is for you by circling a number next to it. Use the scale below to make your choice.

1	2	3	4	5	6	7
never true	**very seldom true**	**seldom true**	**sometimes true**	**frequently true**	**almost always true**	**always true**

1.	My painful experiences and memories make it difficult for me to live a life that I would value.	1	2	3	4	5	6	7
2.	I'm afraid of my feelings.	1	2	3	4	5	6	7
3.	I worry about not being able to control my worries and feelings.	1	2	3	4	5	6	7
4.	My painful memories prevent me from having a fulfilling life.	1	2	3	4	5	6	7
5.	Emotions cause problems in my life.	1	2	3	4	5	6	7
6.	It seems like most people are handling their lives better than I am.	1	2	3	4	5	6	7
7.	Worries get in the way of my success.	1	2	3	4	5	6	7

This is a one-factor measure of psychological inflexibility, or experiential avoidance. Score the scale by summing the seven items. Higher scores equal greater levels of psychological inflexibility.

Bond, F. W., Hayes, S. C., Baer, R. A., Carpenter, K. M., Guenole, N., Orcutt, H. K., Waltz, T., & Zettle, R. D. (in press). Preliminary psychometric properties of the Acceptance and Action Questionnaire – II: A revised measure of psychological inflexibility and experiential avoidance. *Behavior Therapy*.

Appendix 4

CompACT

Please rate the following 23 statements using the scale below:

Strongly disagree **0**	Moderately disagree **1**	Slightly disagree **2**	Neither agree nor disagree **3**	Slightly agree **4**	Moderately agree **5**	Strongly agree **6**

1. I can identify the things that really matter to me in life and pursue them	0	1	2	3	4	5	6
2. One of my big goals is to be free from painful emotions	0	1	2	3	4	5	6
3. I rush through meaningful activities without being really attentive to them	0	1	2	3	4	5	6
4. I try to stay busy to keep thoughts or feelings from coming	0	1	2	3	4	5	6
5. I act in ways that are consistent with how I wish to live my life	0	1	2	3	4	5	6
6. I get so caught up in my thoughts that I am unable to do the things that I most want to do	0	1	2	3	4	5	6
7. I make choices based on what is important to me, even if it is stressful	0	1	2	3	4	5	6
8. I tell myself that I shouldn't have certain thoughts	0	1	2	3	4	5	6
9. I find it difficult to stay focused on what's happening in the present	0	1	2	3	4	5	6
10. I behave in line with my personal values	0	1	2	3	4	5	6
11. I go out of my way to avoid situations that might bring difficult thoughts, feelings, or sensations	0	1	2	3	4	5	6
12. Even when doing the things that matter to me, I find myself doing them without paying attention	0	1	2	3	4	5	6
13. I am willing to fully experience whatever thoughts, feelings and sensations come up for me, without trying to change or defend against them	0	1	2	3	4	5	6
14. I undertake things that are meaningful to me, even when I find it hard to do so	0	1	2	3	4	5	6
15. I work hard to keep out upsetting feelings	0	1	2	3	4	5	6
16. I do jobs or tasks automatically, without being aware of what I'm doing	0	1	2	3	4	5	6
17. I am able to follow my long terms plans including times when progress is slow	0	1	2	3	4	5	6
18. Even when something is important to me, I'll rarely do it if there is a chance it will upset me	0	1	2	3	4	5	6
19. It seems I am "running on automatic" without much awareness of what I'm doing	0	1	2	3	4	5	6
20. Thoughts are just thoughts - they don't control what I do	0	1	2	3	4	5	6
21. My values are really reflected in my behaviour	0	1	2	3	4	5	6
22. I can take thoughts and feelings as they come, without attempting to control or avoid them	0	1	2	3	4	5	6
23. I can keep going with something when it's important to me	0	1	2	3	4	5	6

CompACT Scoring Instructions

Scores are derived by summing responses for each of the three subscales (Openness to Experience; Behavioral Awareness; Valued Action) or the scale as a whole (CompACT Total score).

Twelve items are reverse-scored before summation (items 2, 3, 4, 6, 8, 9, 11, 12, 15, 16, 18, and 19).

Openness to Experience (OE) subscale

- Calculated as the sum of scores for items: 2 (reversed), 4 (reversed), 6 (reversed), 8 (reversed), 11 (reversed), 13, 15 (reversed), 18 (reversed), 20, and 22.
- Subscale scores range from 0-60, with higher scores indicating greater openness to experience (willingness to experience internal events [thoughts, feelings, sensations, etc.] without trying to control or avoid them)

Behavioral Awareness (BA) subscale

- Calculated as the sum of scores for items: 3 (reversed), 9 (reversed), 12 (reversed), 16 (reversed), and 19 (reversed).
- Subscale scores range from 0-30 with higher scores indicating greater behavioral awareness (mindful attention to current actions)

Valued Action (VA) subscale

- Calculated as the sum of scores for items: 1, 5, 7, 10, 14, 17, 21, and 23.
- Subscale scores range from 0-48 with higher scores indicating greater engagement in valued actions (meaningful activity)

CompACT Total

- Calculated as the sum of the three subscale scores, the full-scale CompACT Total score ranges from 0-138, with higher scores indicating greater psychological flexibility.

Reference:

Francis, A. W., Dawson, D. L., & Golijani-Moghaddam, N. (2016). The development and validation of the Comprehensive assessment of Acceptance and Commitment Therapy processes (CompACT). *Journal of Contextual Behavioral Science*, *5*, 134–145. http://doi.org/10.1016/j.jcbs.2016.05.003

Appendix 5

FSHD Values Sort

Instructions: 1) *Carefully read each value and place an X in ONE of the three right-hand columns that* ***best reflects*** *how important each value is to you.* **2)** *Rank your* ***top 5 MOST important values*** *in the MOST IMPORTANT column by numbering them by personal importance e.g. write* ***1*** *for most important,* ***2*** *for next most important, etc…*

Value	Description	Least Important	Somewhat Important	MOST Important
Adventure	Willingness to take risks or to try out new activities, or experiences.			
Advocacy	Actions that influence social decisions especially regarding the needs of those lacking visibility or power.			
Assertiveness	Confident and respectfully forceful behavior to achieve a goal.			
Beauty	Actions that create or lead to the experience of awe or wonder.			
Boldness	Willingness to take risks and act innovatively with confidence or courage.			
Caring	Engaging in kind and supportive actions for others.			
Challenge	Engaging in a task or situation that tests your abilities.			
Compassion	Concern for the suffering of others accompanied by helpful actions.			
Connection	Taking actions that deepen the relationship shared with another person.			
Contribution	Actions taken to advance an important cause.			
Cooperation	Joining with others to accomplish an important goal.			
Courage	Continuing in actions in spite of fear or difficulty.			
Creativity	Actions that turn new and imaginative ideas into reality.			
Curiosity	Actions taken to fulfill a strong desire to know, learn, or understand.			
Encouragement	Actions that provide support, confidence, or hope to others.			
Excitement	Living with great enthusiasm, energy, and eagerness.			
Fairness	Taking actions to promote impartial and just treatment without favoritism or discrimination.			
Faith	Actions that express one's core beliefs both religious and secular.			

Value	Description	Least Important	Somewhat Important	MOST Important
Fitness	Actions promoting one's physical health, strength, and flexibility.			
Flexibility	Willingness to take actions to adapt to changing and sometimes painful circumstances.			
Freedom	Ability to choose to engage in action unhindered by emotion, thought, or circumstance.			
Fun	Engaging in actions that result in enjoyment, amusement, or joy			
Generosity	Giving freely of time, energy, resources, or kindness to others			
Gratitude	To be thankful, ready to show appreciation, and to return kindness to others			
Honesty	Actions that are straightforward, trustworthy, loyal, fair, and sincere			
Humor	The ability to find things funny, and to do things that provoke laughter and joy.			
Humility	Engaging with others without pride or arrogance, valuing the needs of self and others similarly.			
Independence	Being able to take care of yourself without being dependent on another			
Intimacy	Experiencing close, familiar, affectionate and loving relationships with others			
Justice	Taking actions that seek to bring about rightness, fairness for oneself and others			
Kindness	Actions that are gentle, respectful, caring, and helpful toward others without expecting return			
Patience	Capacity to accept delay, challenges, or suffering without getting angry or upset.			
Persistence	Firm or obstinate continuance in a course of action in spite of difficulty or opposition.			
Pleasure	Engaging in actions that generate feelings of happy satisfaction and enjoyment.			
Power	Actions that utilize ability to direct / influence the behavior of others or the course of events.			
Reciprocity	Engaging with others in actions that provide mutual benefit.			
Respect	Actions that regard for the feelings, wishes, rights, or traditions of others.			
Safety	Actions that protect self or others from danger, risk, or injury.			

Value	Description	Least Important	Somewhat Important	MOST Important
Self-care	Practice of taking an active role in protecting one's own health, well-being and happiness.			
Sexuality	Sexual feelings, thoughts, attractions and actions directed towards others.			
Spirituality	Activities that promote connection with that which transcends the self			
Trust	Taking actions that exhibit confidence in the reliability, truth, ability, or strength of others			
Vitality	Taking actions that reflect physical / mental energy or strength, and that nurture life and growth for self and others			

Appendix 6

Fatigue-Related Thoughts Record					
Situation	**Feelings**	**Thoughts**	**Mindfulness**	**Values**	**Actions**

Appendix 7

My Struggle with Pain

Most people with chronic pain have had numerous past treatments attempting to decrease pain’s negative effect.Please everything you can think of- formal and informal you have done to control pain and how they have worked. Please take a few moments and complete the following table.

What have you done to avoid, reduce,or control your pain?	Short-term results on pain	Long-term results on pain	Long-term results on quality of life

Appendix 8

FSHD Sleep Diary

Today's Date	Sample							
What time did you get into bed?	10:15 pm							
What time did you try to go to sleep?	11:00 pm							
How long did it take you to fall asleep?	25 mins							
How many times did you wake up? (not counting the final morning wake up)	3							
In total, how long do these awakenings last?	1 hr 40 mins							
What time was your final morning awakening?	7:20 am							
What time did you get out of bed for the day?	8:30 am							
How would you rate the quality of your sleep? **See below:** Enter the appropriate number.	1 - poor							
Comments: Please note factors that you think may have affected your sleep time and quality. Especially note things like: *Illness, pain, life stress, worries, exercise, use of caffeine, screen time, partner's sleep, etc..* Note when these occurred.	Worries about money, getting a cold							

Sleep quality rating:				
Very poor = 0	**Poor = 1**	**Fair = 2**	**Good = 3**	**Very Good = 4**

Appendix 9

Weight Self-Stigma Questionnaire

Completely Disagree	Somewhat Disagree	Neither Disagree or Agree	Somewhat Agree	Completely Agree
1	**2**	**3**	**4**	**5**

Instructions: Please use the one - five rating scale just above to indicate what level of agreement or disagreement you have with the 12 statements below. Circle the number that best expresses your level of agreement with the statement.

Item	Rating				
1. I'll always go back to being overweight	1	2	3	4	5
2. I caused my weight problems	1	2	3	4	5
3. I feel guilty because of my weight problems	1	2	3	4	5
4. I became overweight because I'm a weak person	1	2	3	4	5
5. I would never have any problems with weight if I were stronger	1	2	3	4	5
6. I don't have enough self-control to maintain a healthy weight	1	2	3	4	5
7. I feel insecure about others' opinions of me	1	2	3	4	5
8. People discriminate against me because I've had weight problems	1	2	3	4	5
9. It's difficult for people who haven't had weight problems to relate to me	1	2	3	4	5
10. Others will think I lack self-control because of my weight problems	1	2	3	4	5
11. People think that I am to blame for my weight problems	1	2	3	4	5
12. Others are ashamed to be around me because of my weight	1	2	3	4	5

WSSQ Scoring Instructions:

Self-devaluation subscale (Items 1-6) Add the rating score for items one – six to calculate the self-devaluation score.
Enacted stigma (Items 7-12) Add the rating score for items seven – 12 to calculate the enacted stigma score.

Total Score: Add the rating score for all twelve items calculate the total score.

Reference

Lillis J, Luoma JB, Levin ME, Hayes SC. Measuring Weight Self-stigma: The Weight Self-stigma Questionnaire. *Obesity*. 2009;18(5):971-976. doi:10.1038/oby.2009.353.