

SYNAPSE

The Newsletter of The Spastic Paraplegia Foundation Inc.

Volume 23, Issue 2

Spring 2020

WHAT'S INSIDE

Resilience Matters Part Three

Page 5

A Career Sidetracked

Page 6

Rare Disease Week on Capitol Hill

Page 7

No, It Wasn't My Worst Nightmare, It was Real

Page 9

Mobility Aids

Page 12

Exercise and Stretching

Page 12

Letter from the President



Frank Davis

Dear Friend,

I always enjoy writing to you for this newsletter and it never seems like we have enough chances to communicate. Well, today is Easter Sunday

and it is an interesting time for our editor, John Staehle, to have asked me for my thoughts. I hope you will forgive me if my rumination sounds too much like a sermon.

Easter and Passover both deal with severe Tragedy, Death and Hope; subjects particularly poignant in our Coronavirus times. The interesting thing about the juxtaposition of all these words is that this is a large reason why many atheists choose to be atheists - there can't be a loving God that allows such tragedy to happen. Many of the smartest minds, in the history of mankind, have for centuries tried to answer this question in sometimes extremely long and complicated books.

This is a time in history when things probably look similarly as bleak as they did to the followers of Jesus when they saw him dying on the cross or when the Jewish people heard the Egyptians marching toward them, facing almost sure massacre. We are all having to stay locked up in our homes while we

hear horrible statistics in the news every day - not knowing when it will end - not knowing who will get sick or die.

The question of "why" is not just a philosophical or religious question, particularly for our community of people suffering from a host of rare diseases. "Why did this happen to me or to my close friend or loved one?", is a question that probably popped into your mind automatically the moment the diagnosis was heard - or even before when you or your loved-one experienced the agonizing, progressive symptoms.

I think the answer is found in the Easter and Passover accounts and it is probably best told in the narratives themselves. We have all heard at least one or the other story so many times that it has become more cultural than directly meaningful as they are meant to be. Like poetry, I think the message can mean more in the metaphor than it can when we try to break it down into a numeric, logical argument, but we really need to pay close attention to the storyline.

The answer, I believe, is that even in the most tragic of times, Love and Hope exist in a grand and momentous form. Love and Hope exist for each of us regardless of who we are or how bad our symptoms have become. When Jesus met the first people that witnessed

Continued on page 3

In This Issue

	Page
President's Letter	1
From the Editor.....	3
QR Code for Ease of Donations!.....	3
About Your <i>Synapse</i> Subscription.....	4

SPF 2020 Virtual Annual Conference

SPF Virtual Annual Conference	4
-------------------------------------	---

General Interest

Resilience Matters - Part Three	5
The Rocky Road to Diagnosis	6
A Career Sidetracked by a VUS.....	6
I had Ataxia for 27 Years	7
Rare Disease Week on Capitol Hill.....	7
My Name is Constance Rucker.....	8
Camping - Part Two	9
No, It Wasn't My Worst Nightmare	9
Chapter Two	10
Notes from Hospital and Rehab Stays..	10
It Needs More Power.....	11

Living with HSP/PLS

Mobility Aids	12
Exercise and Stretching is Key.....	12
My Take on Physical Therapy.....	13

Connections

North Texas Connection.....	14
-----------------------------	----

Other

Let Me Muscle In (by M. Dollinger).....	4
SPF Online Store	15
Loving Better Today (by J. Blaylock)	16
Speech (by H. Fenlon)	16

SYNAPSE

Volume 23, Issue 2 - Spring 2020

The Spastic Paraplegia Foundation Inc. (SPF) is a national, not-for-profit, voluntary organization. It is the only organization in the Americas dedicated to Primary Lateral Sclerosis (PLS) and Hereditary Spastic Paraplegia (HSP).

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President's Letter (continued)

his resurrection, he did not greet them with a lecture. He did not greet them with a rebuke. He said "Greetings. Do not be Afraid!" You don't have to believe in Jesus as a messiah to consider that a wonderful greeting. He did not ask them to pity him nor to strike their enemies. He did not miraculously remove all their problems. He just said, "Do not be Afraid!" That same message is repeated many times in both the old and new testaments. If we have courage and trustingly believe in that hope, a light at the end of our tunnel will enlighten whether or not our physical disease symptoms start to subside.

If we sincerely and patiently believe in that Hope and Love, it will become more and more contagious and synergistic. Great people work together, wonderfully, in response to tragedy. Examples of that can be seen all over the place in the wake of this Coronavirus tragedy. I was in the citrus business and have lived through many hurricanes in both south Texas and Florida. Many of us remember, in 9-11, how people performed together so wonderfully and sacrificially that great outcomes did happen.

You know that the Coronavirus did not stop the need or the mission of our Spastic Paraplegia Foundation. We need to be even more diligent. We need to be even more coordinated and hard working toward meeting our goals of cures for both HSP and PLS! We need to be relentless in our determination. You know that many people in our SPF community have temporarily lost their jobs so the record-breaking giving power we had last year may have quickly dwindled. If that is you, I am very sorry and I hope your employment soon recovers. If that is not you, I hope that I can challenge you to give even more in 2020 to make up for that loss and maintain our momentum.

As I'm sure you know, we were forced to cancel our Annual Conference scheduled for June 25th to the 28th in Nashville, TN due to problems controlling social distancing throughout the 4-day event. Everyone who registered has been notified and refund checks have been issued. We are planning a Virtual Conference online that will begin on June 26th. An agenda will be emailed to all of our SPF community and internet links will be included to easily register to attend at the scheduled times. You can also visit the SPF website, www.spf-foundation.org, to register online now. Most of the same speakers will be speaking online over the next few months. We are arranging opportunities for you to ask questions. Our Conference Manager, Norma Pruitt, is doing an excellent job of arranging and coordinating this Virtual Conference.

Our crucial mission continues! We can do it! I hope you are wholeheartedly joining with me in this effort! I will close my almost unintended sermon today with Psalms 90:17: "Let the favor of the Lord our God be upon us and prosper for us the work of our hands - O prosper the work of our hands".

Frank

Frank Davis, President

QR Code for Ease of Donations!

Using your smart-phone, scan this quick response (QR) code as a convenient tool for submitting donations to the Spastic Paraplegia Foundation. Scan it with your smartphone and a QR reader app and it will go straight to our donate page. Your contribution allows for medical research of Hereditary Spastic Paraplegia (HSP) and Primary Lateral Sclerosis (PLS), together we are #hspandpls.



From the Editor

The Spring issue of *Synapse* was more difficult to prepare than previous issues of the newsletter. Consequently, it is being published a couple of weeks into Summer. Normally, the Spring issue focuses on the Annual Conference including the daily program schedule, the biographies of the featured speakers, hotel reservation information, host city attractions and more. The Covid-19 pandemic changed our normal this year. It created a void in the normal 20-page issue that I couldn't fill. In addition, my editing duties were interrupted by a 2½ month stay in a rehabilitation facility. If you have HSP or PLS, don't break an ankle.

The call for articles for the Summer *Synapse* has already been issued and the newsletter should be published on schedule.

John Staehle, Senior Editor



Normally, the Annual Conference is held the last week of June. The Covid-19 pandemic changed our normal for this year. The Annual Conference, scheduled for June 25th to June 28th in Nashville, was canceled and shortly thereafter, efforts began to organize a virtual conference. The first two speakers made their presentations on June 26th and 27th using Zoom and concurrently live-streamed on YouTube: on Friday evening Corey Braastad, PhD, discussed Basic Genetics and Cell and Gene Therapy Updates and on Saturday morning, John Fink, M.D., SPF's Medical Advisor, discussed the research being done on HSP and PLS.

All the featured speakers from the canceled conference expressed a desire to make their presentations via Zoom and YouTube. SPF's conference coordinator, Norma Pruitt, and her small staff of "techies" are working out the details for scheduling and conducting the next round of presentations. To view SPF and related videos on YouTube, go to YouTube Videos and search spasticparaplegiafoundation. Visit the SPF website, www.spf-foundation.org often for announcements regarding schedules for additional speaker presentations. Also, register for future Zoom presentations on the SPF website. Attendance at the 2020 Virtual Annual Conference, whether by Zoom or YouTube, is FREE.

Online or Printed

When you subscribed to *Synapse*, you chose "Synapse (online version)" or "Synapse (printed copy)" or you may have chosen both.

There are two advantages to reading *Synapse* on the web: (1) it's posted within one day of the editor's approval which is about 5 to 7 days sooner than when mailed copies arrive in mailboxes; and (2) the links throughout the issue are active allowing you to click and go to the site. You can also download the latest issue in PDF format to read offline or print using your printer.

If you currently receive a printed copy of *Synapse* and wish to discontinue it, please go to the SPF website, click on Join Us and Stay Informed, complete the required fields and check just the *Synapse* (online version) box. You will be removed from the mailing list and you will save the SPF \$5 for each issue you no longer receive in the mail.

Let Me Muscle In

"Only a flesh wound"

I remember from childhood

Trivial injury to the good guy

The bad guy gets killed

Flesh is taken for granted

Or rather, muscle

Connected to bones to make things work

Like sitting, standing, walking

Tying my shoes

Getting from here to there

Without a prior business plan

Taken for granted all my life

Now I have a new specialty

Planning my center of gravity

Life is a series of controlled falls

Always something to grab

To stop myself

I dread the moment, someday

When I will miss

Good friends are my lifeline

There are two kinds

Old friends and New friends

The old ones remember me

Before my disability

They see me walking in their memory

Or they don't care about my walking

The new ones don't know me

Except in my wheelchair

What do they think?

Can they imagine how I used to be?

Can a pretty woman look at me as a man?

In fact can I do the same?

Some new friends have a gift

Of not caring about my wheelchair

Or pretending rather well

I need to prove myself

The part of me that's left

My intellectual side

Untouched by my flesh wound

So I keep searching for the clever thought

To make it all better

From "Life is a Journey"

by Malin Roy Dollinger, Xlibris, 2015

Resilience Matters – Part Three

Building Resilience

By Kathi Geisler, President of Life's Still Good



Kathi Geisler



People typically work through three stages to develop greater resilience.

The first is **Gaining Knowledge and Understanding**. This involves investing time and energy researching your condition – the symptoms and how the condition is likely to change over time. It also helps to seek out others with your condition to compare situations and resources.

The second stage is making changes in **Managing Daily Life**. Learn new coping

strategies and lifestyle behaviors to discover how to best take care of yourself physically, socially and financially. Try different methods for managing stress and unpredictability. New, helpful techniques will help build confidence to better handle your situation.

The third is an “acceptance” of your situation, leading to **Growth and Reconciliation**. In this stage, you start to experience shifts in priorities as a result of a more developed understanding of what’s important and what is not. You may begin to take steps to strengthen relationships and feel a sense of gratitude for the positive things in life.

There are numerous lifestyle factors or habits that you can cultivate to support and build resilience. Check out this list to honor what you’re already doing, and note where you can learn and add new strategies:

Maintain strong social connections – Social support reduces depression and will help you manage stress, leading to better physical and psychological health.

Maximize physical wellness – Focus on healthy eating habits, exercise, sleep and symptom management and therapies.

Set realistic goals – Achieving goals helps you feel competent, fulfilled and in control.

Practice gratitude – Recognize and stay aware of the positive things in life.

Nurture positive emotions – Savor humor, hope and optimism when they occur and engage in activities that result in positive emotions.

Allow negative emotions – Feelings, difficult as well as pleasant, are part of being human. Expressing anger, frustration, grief or sadness is healthy, as long as you don’t stay stuck in them.

Use mindfulness and relaxation techniques – Breathing techniques, imagery, progressive muscle relaxation and mindfulness meditation are helpful strategies to reduce stress and anxiety.

Practice forgiveness – Holding on to resentment and bitterness towards people who have hurt you is associated with higher cortisol and blood pressure levels, as well as psychological distress. Forgiveness is a gift you can give to yourself.

Plan for the future – Account for actual needs and limitations for today as well as in the future.

Find a sense of meaning and purpose in life – Having “purpose”, whether through volunteer work, helping others, social activities, relationships or other avenues has physical as well as psychological benefits.

Foster inner peace/spirituality – Involvement in a faith community or other avenues such as being in nature environments can help foster an inner peace.

Learn to tell your story in a different way – Honor the challenges but also what they’ve taught you and what strengths you’ve developed as a result.

Resiliency Quotes:

“A diamond is just a piece of charcoal that handled stress exceptionally well.”

- unknown

“Hardship often prepares an ordinary person for an extraordinary destiny.”

- C.S. Lewis

“The marvelous richness of human experience would lose something of rewarding joy if there were no limitations to overcome. The hilltop hour would not be half so wonderful if there were no dark valleys to traverse.”

- Helen Keller

“It’s not whether you get knocked down, it’s whether you get up.”

- Vince Lombardi

“Adversity has the effect of eliciting talents, which in prosperous circumstances would have lain dormant.”

- Horace

Kathi Geisler is a Co-Founder of the Spastic Paraplegia Foundation and a former Vice President of the SPF. She has HSP and lives in Massachusetts.

The Rocky Road to Diagnosis

A Genetic Test That Took About Two Years

By James A. FitzGibbon, HSP SPG7

The first clue I had that things were not quite right came when people started asking me what was wrong with my right leg, saying it looked as if I was limping. That was in 2010.

The limp gradually became more of a hobble, so eventually I went to the local hospital to get it checked out in 2013. Following a number of MRI scans, it was hinted that my condition might be neurological or genetic. I was advised to get myself checked at a centre that specialized in this sort of illness. We were abroad at the time – in the Middle East.

About a year or so later, we moved back to UK and I consulted the nearest hospital that could guide me to identify this mysterious illness. I was referred to the local hospital that in turn referred me to another hospital to get a genetic test. This was in September 2016. At the same time, I was also referred to a national centre for neurological illness and research in London. I did lumbar punctures, a few more MRIs, an ECG test, a cognitive processing test and the usual physical checks before diagnosing me with Idiopathic Cerebellar Ataxia. The results of the genetic test were still not in by the time I had to go abroad for employment in October 2016.

Unbeknownst to me, results did come from the laboratory in Oxford that November, but, because the report contained erroneous information, huge areas were redacted and it was never shared with me. When I asked the hospital where I had the genetic testing done about any news in June 2017, it seemed that a corrected report had been forgotten about.

In May 2018, I had a major relapse. The following month, I returned to the UK, went to the local Health Centre, was immediately hospitalised and eventually was referred back to the main hospital in London the following month. Apparently, the doctor I normally saw was busy at that time, so I had to see a student doctor, who, upon reading my genetic report, which had miraculously arrived, diagnosed HSP, SPG7 mutation.

This leads me to ponder: if I had had the genetic report in 2016 or, at least, in 2017, could I somehow have prevented HSP being triggered in 2018? I guess I'll never know, but what is for sure, is that the road to my eventual diagnosis, took about eight years!

A Career Sidetracked by A VUS

(Variant of Unknown Significance)

By Peter Slobodnick, HSP

In 2006, I started EMT Basic training. At the same time, I began caring for my father who was terminally ill with metastasized lymphatic cancer that had spread to other parts of his body.

After his passing, I decided to become a Firefighter Paramedic. I earned two Associate of Arts degrees in Fire Technology and was awarded Phi Theta Kappa academic honors. I had an internship with the Sacramento City Fire Department as an EMT-B. It was the training I acquired from this fire house that influenced me to pursue the medical field. The skills, experience and training I acquired from the Station 56 fire house allowed me to gain valuable patient care experience.

I started my paramedic training in Texas near Ft. Worth because the scope of practice for paramedics in Texas was so much broader than it was in California. I gained my training and various skills from the Benbrook Fire Department and in the Huguley Medical Center's Emergency Room. I was licensed as a National Registry EMT Paramedic and soon after, a licensed California Paramedic.

I built my resume in the medical field as an employee of the UC Davis Medical Center (UCDMC) performing a variety of duties for critical and non-critical patients. I was injured about a year after my employment started. I was the sole Emergency Trauma Technician to cover the entire ER Department. I not only had to run from "pod/area" to "pod/area," but I had to get each nurse or doctor any item they asked me to.

The ER Department was staffed with a minimum of five to seven Senior Emergency Department Technicians; however, I was the only Senior ER Trauma Technician that arrived for his shift one day. I learned from the emergency personnel that the technician on the prior shift did not show up either. The exhaustion and repetitive tasks I had to perform for the entire Emergency Room resulted in an injury, causing me to call in as being hurt. I went to my Primary Care Physician at UCDMC, who took me off of full-time duty and put me on "light duty." I began rehabbing my body and regaining my strength with trips to the gym and exercises in the pool.

My PCP could not see or treat me any more since my injury occurred at work. I then started seeing the employee health doctor who told me I would

need surgery to fix my injury, two herniated disks in my neck. This caused me some concern, so I asked to see a neurologist at UCDMC for a second opinion. The neurologist told me that I did not have the issue that the other doctor said I had. This second opinion left me more confused than ever about how to proceed. I regretfully went forward with the corrective surgery, thinking my original symptoms were going to improve.

My existing health issues were compounded by a new issue that presented itself after the surgery: difficulty swallowing after the replacement of the two herniated disks in my cervical vertebra. I learned I had a herniated disk in my lumbar spine, too. That extended my time away from work. I began to use a cane.

I began searching for other less physically demanding work that I could do at the hospital. I applied for each position I found but had few interviews. I wasn't getting any job leads or interest from anyone in the UCDMC system.

After I was married. I fell under my wife's health coverage. I saw a new neurologist who stated that my previous neurologist told him I could have Hereditary Spastic Paraplegia, not Dandy-Walker Syndrome. I was able to get the genetic test in October 2014 to confirm my HSP diagnosis. I have a variant of unknown significance (VUS) on gene FA2H.

I then began to learn as much as I could about Hereditary Spastic Paraplegia and participated in as many clinical trials as I could. It was more of a fact-finding trial as HSP is a rare disease. The latest clinical trial I was in was with Dr. Jacinda Sampson, who told me that my version had not been documented before. I have joined "Patients Like Me" in an attempt to help those with this rare disease and the medical community in general.



I Had Ataxia for 27 Years, but Now...

By Richard Haag, HSP SPG7

I have Hereditary Spastic Paraplegia (HSP).

Until I was 40 in 1988, I was as healthy as anyone can be. I rarely missed a day of work and my most serious illness was the flu. When I turned 40, I noticed a subtle loss of strength in my legs. Our family physician also noticed a slight abnormality in my gait and referred me to a local neurologist who, after tests that included MRIs, CT scans, and a spinal tap, concluded that I had ataxia. He referred me to a neurologist at Indiana University (IU) to get a second opinion. The IU neurologist confirmed the diagnosis of ataxia.

Ataxia is a rare, progressive, genetic disease that causes weakness in the legs, arms, and difficulty in talking. Even after several blood tests for the genes that were suspected to cause ataxia all came back negative I was treated for ataxia anyway. In 2001, my condition had worsened and a baclofen pump was inserted in my abdomen to reduce the amount of stiffness in my legs. The pump, containing a liquid form of baclofen, delivered the drug directly to my spinal cord through a catheter. At the time, we were living near Columbus, Ohio and I was unable to find a local neurologist who specialized in ataxia.

A Florida neurologist, who maintained an ataxia registry, told me the closest physician to me who specialized in ataxia was Dr. Sid Gillman at the University of Michigan in Ann Arbor, Michigan. I made an appointment with him and after the most thorough examination I ever received, he told me "you know you don't have ataxia". I almost fell off my chair. I had been treated for ataxia for 27 years. I had no weakness in my upper extremities and no difficulty with speaking nor with my eyesight. He suspected that I had a paraplegia which only affects the strength and nerves from the hips down. He referred me to Dr. John Fink also at the University of Michigan. I saw Dr. Fink in May, 2015, and after another thorough examination and a battery of blood tests, he confirmed I had HSP, SPG7, a recessive variant of HSP. Luckily for me, the treatments for HSP are the same as for ataxia.

The word, "Hereditary," scared the hell out of me. The SPG7 gene, being recessive, means I didn't pass the disease to my daughters where they would have symptoms, but I did pass the disease to them as carriers.

The bottom line is, that even though I had been misdiagnosed with ataxia from 1988 to 2015, SPG7 didn't cause any new symptoms to be presented.



Rare Disease Week on Capitol Hill

By Tina Croghan, HSP, SPF Board Member, Missouri SPF State Ambassador

It was a cold and rainy morning before dawn.... Sounds like the opening to a mystery novel. Instead, it was actually the beginning of the trek that Tim, Thunder, my sister Mary Kay and I took from St. Louis to Washington, D.C. to be one of 900 individuals who made the journey to advocate on behalf of rare diseases. To be in a

Continued on next page

picture taken on the steps of the US Capitol and be one of 900 — (sigh) — I was just so lucky to have gone.

We all endured the late February wet and cold and the experience, as a whole, was quite moving and very inspirational!

Our first day was dedicated to learning about all of the items in Congress that the rare disease community needs or that are ready to be approved. They include supporting the creation of The Rare Disease Center of Excellence at the U.S. Food and Drug Administration (FDA), enacting the Newborn Screening Saves Lives Reauthorization Act (H.R. 2507 / S. 2158) and supporting increased funding for the FDA's Orphan Drug Products Clinical Trial Grants Program and the National Grants Program (as well as others). Although there are no specific bills for HSP and PLS pending, all rare diseases would benefit from the increased funding of the National Institutes of Health (NIH) and also having all of our legislators participating in the bipartisan Rare Disease Caucus so that we will have our unique/specific concerns voiced. Rare Disease advocacy is not political; it is neither blue nor red, but rather purple.

One of our breakout sessions was getting together with all of the other rare disease advocates from my state, Missouri. I was pleasantly surprised that there were 13 of us in D.C. representing seven different rare diseases. Most in our group were novices at advocating (like me), but three were "old hands" at this! We decided who would be best to speak to each issue the Every Life Foundation (who sponsored this event) wanted us to share with our elected officials. We all had meetings on our schedules with both Missouri Senators, and then meetings with all of the Congressmen/woman from the different areas of the state where we live. We got to meet with the staff members of our legislators who specifically handle healthcare issues and, by telling the stories of our individual rare diseases and how Congress can support us in our fight for a cure, we all felt that our voices had been heard.

I will say that this was a very humbling and inspirational week in Washington, D.C.! I am so glad that I was there and I hope to go again next



In foreground, L to R: Tim Croghan, Thunder, Tina Croghan

year to advocate on behalf of SPF. I encourage other State Ambassadors and general SPF members to do the same. I have made new life-long friends, both in the rare disease community and on Capitol Hill, and I am excited for what the future will bring us!

My Name Is Constance Rucker and This Is My Story

By Connie Rucker



Connie Rucker

In September 2016 I started walking like I had drop foot. At the time I was working in hospice care and I had to do a lot of walking. I was falling a lot; it was becoming harder and harder for me to walk.

I made an appointment with my doctor who referred me to a neurologist. The neurologist ran many tests, including multiple MRIs, an EMG and months of other tests. It was the middle of 2017 and I still did not know what was going on with me.

MY FAITH IN GOD GAVE ME THE STRENGTH I NEEDED

Then in November 2017, I made an appointment at Emory Hospital in Atlanta, GA. The doctor there looked at the MRI and the EMG tests I had brought with me and concluded that it looked to be ALS. I felt like I had been stabbed in the heart and my eyes filled with tears. I didn't want to hear that word, ALS. I had my family with me and we began to pray. My life changed that day.

I was scheduled for another appointment to do more tests in February 2018. It was then I was told that my diagnosis had changed to PLS. I had never in my life heard of PLS, so I researched it to get as much knowledge as I could about the disease. My symptoms continued to progress to the point I had to resign from my job in May. My faith in God gave me the strength I needed on this journey.

It's very hard for me to walk now. I use an electric wheelchair and a cane to get around. My husband, sisters, kids and my church family all help me out. My prayer is that a cure is found for this horrible disease. I would like to thank everyone that supports the cause to find a cure for PLS.

Camping — Part Two

By Jeanette Boyd

In the years before I knew that I have HSP, I backpacked and camped — hiking up intimidating mountains, swatting vainly at no-see-um bugs, hunkering down in a nylon tent and making campfires with my sons.

Needless to say, my HSP symptoms emerged. Later, those manifestations interfered with my daily living. Climbing over tree roots; walking around a tent to secure guy-ropes; and backing out of the range of a snake; all became things of my past. But my love of being in the outdoors was never covered by dust.

Instead, it went another direction. Namely, I still went outside and traveled — but now via a tiny trailer. In essence, my camping took a modest RV route.

Now, instead of shrugging on a backpack, I shuffle on my walker to our teensy trailer. After loading it with bedding, clothes, and simple food, my able-bodied husband and I attach our trailer to our SUV and head out to “camp” in a park or Army Corps of Engineers site in the late-fall-early-winter months.

True, I no longer am able to hike rock-strewn paths to reach extraordinary vistas. True, I no longer enjoy the satisfaction that comes from carrying everything I need on my back. True, I don’t fall asleep at night with the peace that comes from knowing that I had hiked uneventfully through an area that was home to grizzly bears.

I have traded those unique gifts in for things that I CAN do: things like seeing wild herds of protected buffalos from my parked car; things like listening to coyotes calling at desert’s dusk; things like riding my mobility scooter on short asphalt paths to magnificent views; things like watching astonishing films narrated by rangers at Visitor Centers across the nation.

Most importantly, when I was able to hike and tent-camp, I could share the hours with my husband. I can still do that, albeit, some routes take us intermittently along busy highways where I do get irritable and have white knuckles.

I know that I am lucky to have a mate to assist me in coping with HSP. But I also know that coping, even passably well, with HSP has to address both its emotional and physical aspects. Otherwise, the true impact of HSP (and other neurological disorders) is not fully reckoned with.

I INWARDLY DECIDED THAT ONLY MY BODY WAS CRIPPLED; MY MIND WAS NOT...

No, It Wasn’t My Worst Nightmare Ever. It Was Real!

By Malin Dollinger, M.D., HSP SPG4

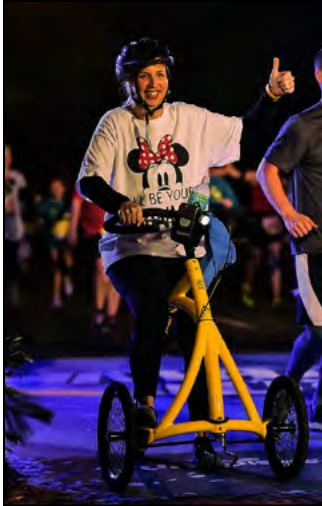
About three months after getting HSP, at age 55, I had an awful thought; I suddenly realized that this was NOT a nightmare. I was not going to wake up soon and realize how awful that nightmare was, and now I was OK; I could walk. Suddenly it dawned on me that I was to be paralyzed for the rest of my life and would never walk again. Or run. Or dance. Or play organ pedals. Or go from here to there without thinking and planning every small part, every step, every weight bearing and movement, every center of gravity, every handhold, every emergency action should I slip and fall. That I would spend the rest of my life as a cripple, a word I’ve never spoken, or written, or used.

I inwardly decided that only my body was crippled; my mind was not, and for the rest of my life I was going to ignore my disability, and/or deal with it so intensively and effectively that overall, it would not make any difference. I could still be a physician, play music, go on trips, be active in organizations, be creative, earn a living, and “pretend” I was just the same as I always was. Now, 28 years have passed since my diagnosis, and I cannot visualize myself in fact being “normal.” My new lifetime normal is now to never walk and to ignore my disability. I do this by inwardly and privately dealing with it every moment of every day and night. Only in occasional real dreams can I suddenly briefly walk unaided, play organ pedals, dance, and get from here to there without holding onto things, I can walk to my car, get in, and drive, can stand up and take things off a tall shelf. These occasional “non-reality” dreams were supported by seeing the opera, “The Death of Klinghoffer,” who appears on stage sitting in his wheelchair after he is killed. He is then able to stand up in front of his wheelchair and walk away. I wondered thus if my soul, as well as his, is not handicapped.

Chapter Two

By Christine Hendrickson, HSP SPG7

My name is Christine. I am 49 years old, I'm a daughter, sister, mother, wife and a nurse practitioner. For years I struggled with fatigue, slurred speech and an unstable gait. I was told it's just depression, it's because you're a busy woman, a single mother of two and your job is demanding. But millions of people deal with the same life challenges, so why were mine so debilitating? After eighteen years of numerous medical professionals, diagnoses, tests and medications and after a neurologist finally recommended genetic testing, I was diagnosed in 2018 with Hereditary Spastic Paraplegia, SPG7, a recessive variant of HSP.



Christine on her Alinker at Disney 5K

Now what? I was a very busy nurse practitioner, a single mom of two teenagers. I went to my neurologist and he suggested that I apply for Social Security Disability Income (SSDI). That rocked my world! I have worked since I was 16 years old. After I got over the shock of this conversation, I filled out the forms for SSDI. In my field I've seen so many people rejected for SSDI benefits (especially the first time they applied), that I was not hopeful. But then I got the call — my application was approved. I was eligible to receive SSDI benefits.

Again, now what? My life as I knew it for 47 years was now going to change.

Time to start my second chapter. My new job was my health. I married a wonderful, caring and understanding man. I gained two stepchildren and both of my children were in college. I started eating better, exercising regularly, sleeping better, volunteering and trying to decrease my stress. One day I saw a YouTube video about Selma Blair and her new walking assistive device, the Alinker. She and I had similar symptoms and this device seemed to improve her life significantly, so I bought one. At first, it was hard for me to master, but I kept trying. Now I can confidently say it has been a game changer. It has given me more confidence and increased my independence. I can do more "normal" things now (and look cool, not drunk!).

In 2021 I will turn 50 years old. I have composed a list of 50 things to do before my 50th birthday. I've become the west Florida ambassador for SPF. I finished a Disney 5K using my amazing Alinker. I volunteer weekly at the VA, donate blood and send packages to the soldiers overseas (my son is serving in the Middle East). Writing this article is another item I can check off the list.



These are just some of the adventures on my 50 before 50 list. You can follow all of them on my Instagram @50.before50.

HSP is still here every day, but I choose to be happy and remain hopeful for a cure.

[Editor's Note: See page 8 of the Fall 2019 issue of Synapse for more on the Alinker.]

Notes from Hospital and Rehab Stays

By Lewis Sid Clark, Illinois SPF Co-Ambassador

I recently was in the hospital ER with stomach pain. This was before coronavirus sent many to the hospital. I ended up having emergency gallbladder surgery the next morning and, with HSP/PLS, rehabilitation at a Skilled Nursing Facility (SNF). Here are some notes you may consider for any stay you may have.

Anesthesia: Due to problems with anesthesia I have had with previous hospital stays, before this surgery I handed out many copies of the article on anesthesia and neuromuscular surgeries (Synapse Spring 2019, pg.14, "Surgical Anesthesia in HSP and PLS"). While the surgical team had a Plan B if things did not go as referenced, it did work and I came out of the anesthesia in good shape. That article is definitely something that should be in your Go Bag. A Go Bag has the many reference material that you will be asked to provide: examples include a list of your doctors, prescription drugs and over-the-counter medications you take, previous operations, contact list, and of course, insurances. For a more detailed list of specific items to put in your Go Bag, see my article, "Go Bag Checklist" in the Winter 2018 Synapse, page 8.

Drugs: For the complete list of my drugs, I showed dosages, prescribing doctor and timing. I gave it to the ER on admittance. Word of warning, you need to be responsible and watch what the

hospital actually gives you. They took my list and merged it with what they had on file from years' earlier visits! So, I was warned some of the drugs showed duplicated treatments. They ignored the timing and, for example, simply wanted to give me all my Bacofen at one time instead of spaced out over a 24-hour period. It took a visit from their Pharmacist to straighten things out. Be alert, check the drugs and dosages they are giving you every time they bring you your meds.

Care Team: While in the SNF for rehab, I had a variety Doctors, RNs, CNAs (Certified Nursing Assistant) and PCT's (Patient Care Technician) all of whom work various days, times and shifts. While the ones that are assigned to you any one day may change, learn the names of those that help you the most. Learn shift change times since it is hard to get service then, anticipate and ask for help before then. Remember they are not just outside your door waiting for your call button. It takes time for them to react and you need to know what an emergency is and what is a "nice to have." It is also a nice gesture to turn in "Super Star" recognition for exceptional service. If needed, file a complaint form when there are unresolved continuing problems.

Call Button: I always had my iPhone within reach and charged. More than once, I had call button problems, especially in the SNF. The call button was left out of my reach. Also, I have had call button failure! The button did not work! Since I marked down the nurses' station phone number and had my cell phone, I called them for help. Yes, they were surprised to have a patient call them, but I got the help I needed.

Go Home: The goal of any stay is for you to be as independent as you can be and for you to go home. Discharge is a function of your medicine, condition and insurance allowances. If you did some therapy but it was not recorded, your records indicate you did not do it. Have any, and all, efforts and improvements recorded. Take every opportunity for Physical Therapy and Occupational Therapy that is offered. After I was home, I had in-home PT/OT followed by outpatient rehabilitation. Medicare and private insurance companies set limits on the amount that is covered. Do additional exercises on your own and follow through on the suggestions offered. Keep all your discharge papers and note any follow up appointments needed and referrals.



It Needs More Power

By Kyle Alumbaugh, HSP

Having HSP since birth has had its challenges. My father and brother also have this disease. Growing up, fear of rejection and failure were two key challenges. As I grew older, the degenerative attributes of the disease started making it more difficult to walk.

As I was pursuing my degree in my early 30s, I was falling very often on and off campus with the assistance of a cane. My wife and I had decided that a wheelchair was the next step. Through this transition, we had help from Vocational Rehabilitation Services. Any of you readers who are pursuing employment or a degree, please seek help from Vocational Rehab in your area. They have helped with a home remodel for my wheelchair, vehicle modifications, funding for college and more. In 2018, I discovered that pain in my arms was gradually coming in. I imagine some of you may have had this problem, too.

Through my wheelchair vendor, I discovered an add-on to my manual wheelchair that gives it power. It's called Smart Drive MX2+ and it is made by permobil (for more information, go to <https://permobilus.com/products/tilite-manual-wheelchairs-smartdrive-power-assist/smart-drive/>). It is a motorized power assist solution for its users that bridges the gap between power and manual wheelchairs. It responds to a Bluetooth connection a smart watch, the Push Tracker also from permobil. I tap my wrist twice and the chair moves. You can set acceleration and speed in 10% intervals. It's been really great



for getting around in the community. It's made my life much easier and allowed me to be more active. It allows me to battle through fear of failure and rejection with great confidence.

Readers, fellow owners of this long, life grabbing disease, hold on. Sure, we get down, upset, angry at the challenges among us. Yes, these challenges are our own, but the challenges of life take hold in many forms. Whether you get a flat tire, the dog poops on the floor, your wheelchair breaks, kids track in mud on fresh clean floors, your feet are super sensitive causing pain or mom becomes critically ill, we can take joy and persevere. Each challenge has a different approach. Plan for the win and tackle life with joyful courage! This isn't our end, folks. This isn't the red light. So, GO!

Mobility Aids That Gave Back My Independence

By Marlene Doolen, HSP, SPF State Ambassador - Central Texas Region

In my mid-40's, I began to experience walking and balance problems. Knowing a relative of mine had a rare disorder, Hereditary Spastic Paraplegia (HSP), I felt I might have HSP, too. I made an appointment with a neurologist who diagnosed me to have HSP. The doctor suggested I, "use a cane so people will not think you are drunk." After leaving the doctor's office, I cried and decided to do my best with what I had or could have. Then I began to use a cane.

I started to observe my feet were not moving well when driving. So, I decided to investigate adding hand controls to my vehicle, a Honda Odyssey minivan. Once I knew that was what I needed to do, I got some education and training on how to drive safely with hand controls. It was offered by a hospital in Austin, TX because they had a car with hand controls for training purposes. An instructor rode in the car, with me driving, to teach me how to use the hand controls. I had to have a couple of times to get through the process of driving in the area around Austin downtown. Insurance didn't cover the cost of the training so I had to pay the hospital fee out of my pocket.



Hand Controls

I was convinced I could do it, so I had hand controls installed on my Odyssey. Before I could get a revised Texas driver's license with a restriction that allowed me to drive only vehicles with hand controls, I had to prove to the Texas Department of Motor Vehicles (DMV) I could

drive safely with hand controls. I demonstrated to a DMV evaluator I knew how to operate my vehicle using the hand controls before I was approved to drive that way. I received my new driver's license and was issued a handicap tag to hang on the rearview mirror when I drive and when I park in spaces designated for the handicapped.

I started using a walker for balance as the tips of the soles of my shoes were wearing away. I only used the walker for short distances and

in the house. For longer distances, I purchased my first 4-wheel scooter. I also purchased a Bruno Joey lift that was installed in the rear area of my Odyssey. I kept the scooter in the back of the minivan to use



Joey Lift inside the rear area of the Odyssey



Folded Walker in the rear seat

when I went shopping. The walker was placed in the back seat of the minivan to use when I had to get from the driver's seat to the rear of the minivan where the scooter was located.

When I began to have more difficulty getting around in my home and the 4-wheel scooter couldn't get through the doorways, I purchased a more maneuverable 3-wheeled scooter to travel around the house. It let me continue to safely get most household things done. When we moved into a new more accessible home in 2010, I decided to purchase a new 4-wheel scooter for shopping. I gave my first 4-wheel scooter to a handicapped Veteran.

A couple of summers ago, my son and his family came from Virginia to visit us. We started at one end of the mall and went to a "Texas" store to purchase some Texas-related items. I was leading the way "driving" my scooter through the mall. All of a sudden, I heard my son hollering, "Hey, Mom, you're way ahead of us!!" We all had a good laugh and then I slowed down to let them catch up.

Exercise and Stretching Is Key

By John Boucher HSP, SPG7

Following a routine exercise and stretching program is crucial for the Hereditary Spastic Paraplegia (HSP) patient to function optimally.

As we all are aware, HSP makes muscles malfunction causing cramping and muscle spasms. HSP is a large group (currently around 80 types) that make walking difficult due to

weakness and spasticity in the legs. The 80+/- types of HSP are further divided into pure and complex forms. Pure primarily affects the lower extremities and back. Complex involves the lower extremities and back as well as the arms, eyes, throat and possibly the cerebellum. The primary problem is that the signals our neurological system sends to various parts of our bodies do not reach their intended destination. Currently there is no cure for this.

Since there is not much we can do about the neurological system, but there are things we can do to maintain muscle function. The human body really does amazing things and can figure out how to work around problems. Just walking is difficult, but if we continue to push and fight, our body will figure out how to make the required muscles work in "nontraditional" ways. Often times this requires a little extra work and requires one to be cognizant of simple movements. For example, we have to concentrate on walking properly by having the heel strike the ground first, fighting against foot drop. You should start any program like this slowly and cautiously in a safe environment. Perhaps exercise every other day at first and slowly progress to exercising every day. I would recommend stretching 3-4 times a day, every day. Keep a log to track your progress. It can be hand-written or there are some good free apps available for your smart phone like MapMyWalk by Under Armour (it tracks more than just walks).

The most important thing is safety. If you have to use a cane, walker or rollator, so be it. You may ache and feel tired, especially at first, but I would encourage you to push through it and don't give up. Listen to your body, if you are hurt, stop; you know your body better than anyone.

The reason I like to stretch and exercise is twofold – it makes me feel good and hopefully, I can slow HSP's progression until a cure can be found.

Stay positive and keep in mind that the glass is half full! We are not given things we can't handle!



My Take on Physical Therapy

By Helen Smith, HSP SPG4

We do not ignore physical therapy out of lack of time or having better things to do. I have taken long breaks because of the pain it causes. I tell myself - it will keep me more limber and stronger, but it's a challenge.

I started working out again at Magee Rehab's satellite office close to me. The girls there are wonderful. They show this in their interactions with all of us and their observations while we did the exercises they taught us. By the time I finished that round of therapy I felt so much more confident. After my insurance coverage ended, I started what they call Wellness Therapy. For a low cost, I could still go and do what they taught me on their equipment while they were there. I could ask questions as needed, too.

A few months ago, it got to the point where I was telling a friend about the pain I was in and she insisted I think about giving up the PT. So, I talked to my physical therapist. She told me the pain I was having from overdoing it should only last two days. The pain felt like tingles shooting down my legs. Then she recommended I talk to the doctor who prescribed gabapentin for me. I did and he increased the dosage which took care of the discomfort I was experiencing.

I was going regularly but was wiped out the next day. So, I reduced the time I spent using the Nu-Step Recumbent Cross Trainer (nustep.com) from twenty minutes to fifteen minutes. The Nu-Step is a device like



Nu-Step Recumbent Cross-Trainer

a recumbent bike, but instead of pedaling it, you push down on flat pedals and your legs go back and forth instead of around and around. It is my favorite piece of equipment. It not only strengthens my calf muscles but also my stomach muscles. After that I use steps with a railing to hold while I stretch my legs and feet.



Total Gym-Fit

Then I use the Total Gym (totalgymdirect.com). On this, I lie flat on my back at an angle with my legs in a squat position. Using both of my legs, I push my body up the incline until they are straight. Then I return to the squat position and repeat. After using both legs, I do each leg individually. Increasing the angle of incline increases the amount of my body weight I have to push up the incline.

Following that I head to the parallel bars. I stand on a square foamy block and, with my feet next to each other, I lift my arms to test my balance.

Continued on next page

Then I stagger my feet and lift my arms and finish with one foot in front of the other. This last position is the biggest challenge. At first, I could maintain my balance just a few seconds. As I have continued to work on my balance, I now can hold that position for about 30 seconds. The whole time I am doing this I am between the parallel bars which I use to catch myself if I lose my balance. I decreased all the mat exercises I do from twenty to fifteen times each. I can honestly say that when I left the facility, I felt like I had had a workout but I was pain free that day and the next.

The point I most want to make is that I have been doing this consistently for almost a year. It is the longest stretch of time that I have

done physical therapy. I promise you it is worth it physically and emotionally. So, get to work! You'll be so glad you did.

*Note: Since writing this, we were hit with the Covid-19 pandemic. Consequently, I cannot go to Magee Rehab for my wellness therapy. I got a bit lax, but I do have a treadmill at home that I use instead of the Nu-Step. I bought a sponge block that I use to do the balance exercises. The mat exercises I just do on the floor. I haven't been doing them twice a week as before, but I promised myself that I will start. This health crisis has given us all a reason to step back and re-examine life. I am an optimistic person and I feel like we will all get through this. **I wish you all the best and Stay Well!!!***

CONNECTIONS

North Texas Connection, March 7, 2020

By Celyna Rackov, HSP SPG4, SPF Ambassador North Texas Region

SPF members in the North Texas region held its first 2020 connection event Saturday, March 7th; it was lunch at Michael's Italian Kitchen Restaurant in Irving. Our meeting lasted two hours. Members Tina Curfman (HSP), Lenny Piepho (SPG7), Jeana Fraser (SPG4) joined by her husband David Fraser, and Celyna Rackov (SPG4) joined by her husband Randy Rackov participated.

The meeting began with introductions and social conversation to get to know one another a little better. Next, members shared information about their diagnostic onset age. Celyna began having symptoms as a child, Tina's symptoms began in her teen years while Jeana and Lenny did not have symptoms until adulthood.

Conversation then shifted toward rare disease day as it was the Saturday prior and fresh in everyone's mind. We learned that there are more than 7,000 known rare diseases and 1 out of 10 Americans has some type of rare disease. We concluded that having a rare disease may not be so rare. With so many rare diseases, it is important that we increase public awareness of both HSP and PLS as so many people with these diseases often are initially misdiagnosed.

We moved on to talk about supplements like Vitamin D, magnesium, and a few others as well

as antioxidants. Tina spoke about her article on the Winter Synapse, *Staying Active with HSP*. Inspired by her speaking, each one of us then began sharing what we do to keep active. Activities such as Pilates, hydrotherapy, water work out, golf, tennis and stretching were all discussed at length.

Walking aids, such as canes, crutches, walking sticks and leg braces were next in our discussion. One aspect of using these aids is the differences in using the aids on both sides or just on one side. It seems to be just a matter of preference;

however, this type of thing should always be reviewed with a licensed therapist so as not to cause a new problem from favoring one side over the other.

Lastly, we spent some time lamenting over disabled parking availability. Disabled parking is often legislated requiring a minimum number of disabled spaces. Many

businesses treat disabled parking places as a waste of space providing the bare minimum and occasionally not placing them in an optimal place.

At the time, there was no Stay-at-Home order and no coronavirus cases in the area, but we were already worried about COVID-19, as people with complicated HSP and PLS are in the high-risk group. We had scheduled our next get-together for May 2nd in case everything would be back to normal. Sadly, that date will have to be pushed out.



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***trotting on new ground
with my black brothers,***

***we stand for togetherness,
the same blood in our veins***

***the future lies open before us
and we step beyond old pains***

***forgive us our trespasses I
say as I wave goodbye to***

***the savagery and ancient
ways of the darker days***

***shame, nor anger, are now
allowed to hinder further...***

***let's turn the pages on
those most cruel ages***

***not forgetting yesterday
but loving better today***

***james kenneth blaylock
6-15-20***

SPEECH

I HAVE A SCLEROSIS THAT'S REALLY ATROCIOUS
IT MAKES ME SPEAK SLOWLY AND SLURRED
I MESS UP MY ESSES SO PEOPLE MAKE GUESSES
TO TRY TO DISCERN MY WISE WORDS
BUT THOUGH I STILL MUMBLE
MY THOUGHTS AREN'T A JUMBLE
IN FACT THEY'RE THE LOVELIEST VERSE
FOR THOUGH I COULD GRIPE
AND SPIT OUT SOME HYPE
IT NEVER SEEMS USEFUL TO CURSE
BUT CURSE I STILL DO
LIKE WHEN SOUNDING THE FOOL
WITHOUT EVEN DRINKING A NIP
FOR THOUGH I DO MUMBLE
AND SOMETIMES JUST GRUMBLE
YOU'D BE WISE IF YOU DON'T READ MY LIPS!

HUGH FENLON, 2018