

SYNAPSE

The Newsletter of The Spastic Paraplegia Foundation Inc.

Volume 24, Issue 3

Summer 2021

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Letter from the President



Frank Davis

Dear Friend,

The mention of summer often conjures images of vacations and a welcomed change of pace. For the Spastic Paraplegia

Foundation, though, there is no summer break. Historically, summer for SPF is one of the busiest seasons of the year. This year is no different.

As I write this column, we have just held the first of the Spastic Paraplegia Foundation's 2021 Annual Conference

Online Series. In 2020, due to the pandemic, we had to swiftly pivot from our traditional in-person Annual Conference to an online event. The experience gained last year allowed us to expand this year's event, resulting in a virtual conference series of four presentations from June to August.

Dr. P. Hande Ozdinler, Associate Professor in the Department of Neurology, Northwestern University, Feinberg School of Medicine, launched the series with

a fascinating look into the world of Upper Motor Neurons (UMN): what they are, what are the roadblocks to having effective treatments, how to overcome those barriers, and the path toward curing UMN diseases such as ALS, PLS and HSP. Dr. Ozdinler's extraordinary ability to explain complex information in an approachable, understandable manner made this session a successful start to the 2021 virtual conference. Her full SPF presentation is available on the SPF YouTube channel. By the time this edition of the

Synapse reaches you, videos of the series' other leading experts in HSP and PLS will be posted, as well. I encourage you to

check it out.

The 2021 HSP and PLS Awareness week is August 21-28. The theme this year is "Challenge" and I challenge you to help raise awareness of HSP and PLS. Share your story and support by participating in one of the special Awareness Week virtual events highlighted on page 13. Awareness Week is also a great time for fundraising and friendraising! Consider organizing a Virtual 5K Run, Walk or Roll. Recruit

"TOGETHER, WE ARE AN UNSTOPPABLE FORCE FOR GOOD!"

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Special thanks to Gulf Business Printing, Jim Burkhalter, and Hank Chiuppi for their assistance and support for the Summer 2021 issue of Synapse.

SYNAPSE APPEAL

Synapse is an online and print publication for members and friends of the Spastic Paraplegia Foundation. If you or a loved one enjoyed this issue and would like to support it, please use the enclosed response envelope or go online at www.sp-foundation.org to make a donation.

EVERY LITTLE BIT HELPS.

SYNAPSE

Volume 24, Issue 3 - Summer 2021

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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Synapse is published quarterly. It is also available online at www.sp-foundation.org

The SPF is a non-profit 501(c)3.

Tax ID # 04-3594491

Combined Federal Campaign CFC #12554

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family and friends to participate. The average person can walk 3.1 miles in about an hour. Use creative calculating and count your time spent stretching, exercising, or with a physical therapist, toward your progress in a Virtual 5K. Invite family and friends to sponsor your FUNraising 5K activities.

The dollars you raise are essential to funding research to find a cure for HSP and PLS. Because of YOU and the leading edge research you make possible through your donations, 135,000 people throughout the world face the prospect of a brighter, more mobile life with reduced pain in the next five to ten years.

You can learn more about the excellent and very promising research and innovation your donations make possible in the *Annual Gratuity Report* available online at sp-foundation.org. This report is another of our summer's priorities.

I cannot say "thank you" enough for the gifts, dedication and efforts of SPF members and friends. Together, we are an unstoppable force for good!

Sincerely,

Frank

Frank Davis, President

URGENT! URGENT! SPF Needs Your Assistance.



The Combined Federal Campaign or CFC is one of the largest fundraising campaigns in America. It is available for all Federal employees.

Starting September 1st, Federal employees can select which charities they would like to contribute to for the following year.

Do you know any Federal employees? They include postal workers, military personnel, police, and many others. If you do, please reach out to them. Let them know about your disease and the work SPF is doing to find a cure. Then ask them to choose Spastic Paraplegia Foundation during the selection process this Fall. **Our CFC number is 12554.**

Please help us with this great opportunity. We challenge each of you to recruit and get a commitment from at least one Federal employee.

HOW TO HELP

We operate out of the strength of our community, caring friends and sponsors. Your help makes a difference!

Please contact us at volunteer@sp-foundation.org to help in one of the areas below or to suggest another way you can get involved.

SUPPORT RESEARCH TO SPEED OUR CURES BY VOLUNTEERING

Below you'll find information on some of the ways you can help SP Foundation in their search for a cure to PLS and HSP.

Raise Funds: The primary focus of SPF is to raise funds to support research to find the causes, treatments and cures for Hereditary Spastic Paraplegia and Primary Lateral Sclerosis. Our major fundraising activity consists of a TeamWalk. Individuals can help organize local fundraisers. People are also needed to secure corporate sponsorships and help with grant applications.

Patient Connection Programs: Organizing a Connections gathering for people to meet, share stories and help one another is a great service. Events can be as simple as meeting for coffee! In areas with large patient populations, SPF seeks to establish Chapters.

Conference Organizers: SPF's conference coordinator gladly welcomes planning and organizing assistance from SPF members living in or near the metropolitan areas selected for annual conferences. These events feature speakers and programs on special topics of interest to our community and provide opportunities for individuals to meet others. Conferences can be half-day or full-day events.

Communications: Individuals with writing, research, website or graphic design skills are needed to assist with various communications initiatives.

Ambassadors: Ambassadors raise awareness about our disorders as well as enhance community building and industry relationships. You can assist with media relations, share your story, speak at local groups or help with grassroots advocacy.

Business and Administrative Support: Volunteers with business and administrative skills can play a valuable role in administering the work of the SPF. Most of the help is coordinated through email correspondence and uses popular Office applications.

Life in a Triple Convergence

By David Buseck, PLS

One day in March 2021, as we were starting to see an end to the pandemic, someone asked me what my ideal first gathering would look like. As I thought about this question, it became obvious to me that I have no clear answer. First, I have to figure out how to navigate the understandably evolving guidance from health authorities about COVID-19 protections as more people get vaccinated. Second, COVID's start coincided with two other significant life events for me:

- In April 2019 I was diagnosed with PLS or HSP, which was refined by August into a more solid PLS diagnosis, and
- In September 2019 I retired from my career of 32 years.

My first six months of retirement were much as I had planned, with lots of music and travel. I took 10 trips that included visits to a new country, a new baby, a funeral, hiking in the mountains, a music festival, family and friends around the U.S. I also got to celebrate a major professional award that was granted to my father, who will probably never retire.

In a normal year, I would have continued to use my new-found time in retirement for more traveling, plus socializing and increasing my volunteer work in the community. I would have been concurrently evaluating each activity with respect to my post-diagnosis awareness of reducing mobility and needed accommodations. It would have been a year of organic growth in terms of understanding how much I can do, what works for me, and what I might need to adapt.

Instead, I established a COVID rhythm that is much slower and smaller than I ever could have imagined. Fortunately, staying home is not too bad for an introvert like me who likes reading, writing, hiking, playing music, and spending time with my favorite person. Each of these activities can be done alone or with my favorite person in the world, so this year has been good for that. My small accommodations include sitting more than I stand while playing

my flute, using a cane on most outings, and using walking sticks on hikes. I also carry a campstool for rest breaks and pick shorter trails.

Traveling was part of my retirement agenda, but of course COVID squashed those plans. I cancelled at least eight trips because of the

pandemic, including Nashville for my first SPF conference. I would have expected this mass cancellation to feel devastating, but it didn't. Much as I had looked forward to each of those expeditions, the totality of them was actually getting pretty overwhelming. I felt my stress go down and breathed easier when there was less on my calendar.

At the same time, some stress increased since traveling is now more of a race against time than I had thought it would be before diagnosis. We were already planning on front-loading our more adventurous trips since we cannot know how age will slow us down, but that urgency is now accelerated. Unfortunately, PLS didn't have the courtesy of pausing its progression during this time of travel restrictions. I don't know how quickly or how far my disease will progress in the future, nor the ways in which it will affect my walking and other functions. Will I reach a point where it feels too difficult to travel? How will it affect my feelings of independence and my psyche if I request a wheelchair at the airport to save walking time for things that matter? How will I balance my physical needs against the needs and expectations of travel companions and the people with whom I'm visiting?



Although the pandemic put Dave's new-found retirement plans on hold, his love of music brightened his quarantine routine.

The biggest losses from this travel hiatus were missing visits to new babies and not being able to mourn deaths together with loved ones. These visits of joy or condolence are important to celebrating the paths of life, and all had to be delayed. Now as we seem to be on the upslope, I eagerly look forward to seeing loved ones again. I also wonder how they will react after not seeing me for 18 months or more. Will my cane shock them? I'm somewhat self-conscious about my reduced stamina and my extra need for downtime, but will other people notice?

Both close to home and on the road, I relish a return to small gatherings and meals together. I look forward to hosting people and sharing music at our new single-story house. But I feel uneasy about bigger gatherings. The typical party conversation—standing together while catching up with an old friend or learning about someone new—will be challenging because my legs get tired when I stand for very long. It feels rude to slide into a chair at every opportunity, but that is what works best in my current world. Sitting also moves me further from a standing conversation companion, making it harder for me to hear and forcing me to work my vocal cords more to be heard above the din.

Like most people, I'm navigating the new world of pandemic emergence as we all figure out what is comfortable for each of us. I am both excited and nervous about returning to a normal life, while knowing that my retirement and my disease progression will leave me at a different "normal" than the one I left behind in March 2020.



Logan's Story

By Kristi Kurtz

Logan is a three-year-old boy from central Minnesota who was diagnosed with HSP SPG4 in August 2019.

At ten months of age Logan stopped doing some of the things he was once doing independently, such as sitting and bouncing in his baby jumper. At twelve months of age Logan began "army crawling" by using his upper extremities to pull his weight around. His legs tried to follow that pattern, however they were too weak and tight.

At 14 months old his primary doctor referred us to a pediatric neurologist and a team of physicians at a gross motor delay clinic an hour and a half away.

After normal blood testing, x-ray of his spine, hips and pelvis, and two MRIs of his spine and head, the next step was genetic testing. We met with a geneticist and had Logan's blood sent to a lab in California for genetic testing. We had a confirmed diagnosis within four weeks.

As devastating as a diagnosis can be, especially for a parent of a very young child, it meant moving forward and gathering what was needed to provide Logan with adequate equipment, medication, services and support.

Later this year we will be traveling 1,073 miles to Dallas, Texas to establish care with another group of physicians who specialize in pediatric patients with HSP. They will collaborate care with his local team of physicians. They are also involved in HSP research which is very important to us. We know what we know about HSP now from others who were willing to be involved in research.

Logan routinely sees a physical medicine and rehabilitation doctor, neurologist, and orthopedic specialist. He goes to physical therapy twice per week, recently beginning aqua therapy in a warm heated pool at 95 degrees. He takes oral baclofen three times a day. He wears bilateral AFOs (ankle foot orthosis) to help support his weak pronating ankles. Every twelve weeks he gets various Botox injections in his legs in addition to phenol injections every six months under general anesthesia.

Despite the challenges Logan has faced and will continue to face as he grows, he has a heart of gold.



Logan and mom Kristi enjoying the wading pool

Continued on next page

He is an incredibly happy little boy who never fails to make those around him smile with his contagious giggle.

Logan enjoys playing in his sandbox, playing in the water with his little sister, drawing with chalk outside, anything to do with cars, buses or trucks, playing Magnatiles, baking muffins and cookies with his Dad or snuggling with his Mom.

We have loved learning from others' stories and connecting with other families around the country with children who have a diagnosis of HSP; especially, my dear friend Amy from New York whose little boy has HSP SPG4. I met Amy through a Parents of Disabled Children support group on Facebook; she has given Logan and me so much love and support and we couldn't be happier to be on this journey together.

As a nurse myself, I had a patient last year tell me about a genetic condition they have. They shared with me "you can't miss what you don't know." I held onto this because with Logan's limitations, he personally doesn't know any different and lives his life the way he was meant to.



Resignify

By Michelle L. Detoni Coleta, HSP SPG8 VUS



Michelle L.
Detoni Coleta

By definition, resignify means to attribute a new meaning to something or to give a different meaning to what was originally planned. This term summarizes my life trajectory. Well, are you curious?

My name is Michelle. I'm 38, I'm married and I have a 5-year-old daughter. I currently live in the state of Minas Gerais, Brazil. Ever since I was a little girl, I always enjoyed studying and dreamed of being a scientist. I saw myself helping people through research involving the cure of some disease.

For this to be possible, I became a biologist. I did my Master's Degree and PhD in Biological Sciences at the Federal University of Juiz de

Fora. It was 15 years of dedication within the laboratory. I acquired experience in cell biology, biochemistry, immunology, genetics and biotechnology. It seemed that everything went as planned: I was teaching and working in research at the university, I had married and was expecting my little princess. I was living the dream!

But, something unexpected came up.

Shortly before the birth of my daughter, my mother, who had difficulty with locomotion for about 10 years, was diagnosed with a rare disease called Hereditary Spastic Paraparesis (HSP). After my daughter was born, we moved to another city. Our family's routine changed due to the care of my little girl and my mother, whose HSP was progressing.

Also, I noticed that in those two years, the symptoms of HSP were beginning to manifest in me and the symptoms were intensifying very quickly compared to the progression of the disease in my mother. Perhaps this was because I pushed myself excessively in caring for my family and was not listening to the limits of my own body. In this period there were also some emotional problems that may have contributed to the advance of HSP.

At the beginning of 2017 I was diagnosed with HSP; I am the third in my family to receive this diagnosis. Other family members (great-grandmother and two great-uncles) had already presented gait difficulties, but never knew the cause. In 2020 my genetic test showed I have SPG8 VUS.

The pain, tiredness and difficulty walking began to be part of my daily life. Gradually, I realized that I could no longer perform in my profession the way I had always dreamed. The advance of the disease caused depression in me. My dream of working for science was getting further and further away. All my studies, all my effort, all the knowledge acquired and the love I had for science, would serve no use. I couldn't find peace!

I needed to do something to find answers for the questions bouncing around in my mind. I began looking for people with HSP using social media. I soon approached three extremely special people: Jailson Mouzinho, Rejane Mota and Celyna Rackov. We organized, and together with other families affected by HSP, we founded ASPEH Brasil (now known as



Post-conference gathering of Medical Doctors, Symposium Committee (in orange) and relatives

ASPEC Brasil), the first association in Latin America to support people with HSP and their families. In December 2020 we expanded our scope to families affected by Stiff-person syndrome, PLS, Sjogren-Larsson Syndrome and tropical spastic paraplegia.

ASPEH Brazil/ASPEC Brazil brought me closer to the world of rare diseases, many of which I was not previously aware. I heard about various chronic diseases and difficult-to-deal-with diseases and have had the privilege of meeting fantastic people resulting in close friendships. But I still missed using science to do something useful for society and depression was always prowling, chipping away at my focus.

Then I learned of the struggle of the children with Edwards Syndrome (trisomy 18) who fight daily for life. For many years, trisomy 18 has been defined as being incompatible with life, but with grit and determination, children are showing that they can live and be happy with the support of their families.

I realized that I was foolish in my self-pity and that I should be grateful for all that I have. It was a very humbling experience.

My friend Marília C. Branco, president of the Associação Síndrome do Amor, showed me that in order to make a change in life, such as the one our children with trisomy 18 perform, we must first change the way we look at situations. Thus, I started a therapeutic process of resignification and saw that my limitations did not affect what I loved so much: the understanding of science. While living with "rare condition" families, I realized that there are so many scientific terms thrown about that are very difficult for the average person to understand. There was a great need

for someone to explain these terms in a way people could understand so that it would be more useful to them.

I didn't even stop to think. I put all my knowledge at people's disposal as I always dreamed. I did not bring the cure for diseases as was my childhood dream; but, I was able to put all my education and scientific knowledge to good use helping people understand better about the rare disease that affects their family. I was able to give them a new perspective and a renewed hope of a better life, bringing clarity to what was obfuscated.

Thus, in social networks, the Ciência RARA (Science for Rares) Project was born, which is available to families affected with rare diseases for any clarification they need related to the science. Now, with the support of the Instituto Vidas Raras, an association that welcomes families with various rare diseases throughout Brazil, we will be able to make this knowledge reach an increasing number of people.

Try to cast a new look at situations that seem difficult and you, too, can resignify your life!

QR Code for Ease of Donations!

Using your smartphone, 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.



No Stopping Me Now!

By Molly St. James, HSP

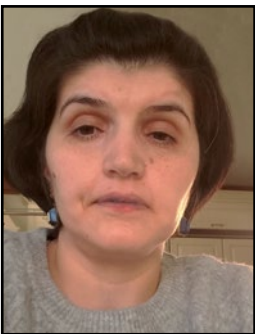
I got walking sticks (www.urbanpoling.com) from my physical therapist; they are awesome. The sticks feel like they remove about 25% of my body weight when walking. Moving about is so much easier! I have only had them a short time so I'm still getting used to them. So far, I only use them when I'm walking outside. When I'm walking inside without the poles, I just keep repeating, "Heel, toe. Heel, toe." On my walk this morning, I was able to wear regular sneakers and I didn't drag my toe once. I feel like I just won the lottery!



Molly and her new walking sticks.

A Journey of a Thousand Miles

By Sofia Weiss, HSP



Sofia Weiss

My story starts when I was ten years old. At school, I noticed that I was not able to jump or climb the stairs as fast as my friends. I knew there was something different about me, so doctors were consulted - many doctors! It was only during my teenage years that they diagnosed me with Spinal Muscular Atrophy (type3). For almost twenty years I have lived with SMA as my diagnosis. In my everyday life, that meant weakness, fatigue, physical pain, and respiratory troubles.

Each night, I use a BIPAP machine. It is meant to assist with my breathing, but it dries out my mouth; it's a balance. My bed is electric, so my back is supported and my head stays elevated, which helps with my breathing and even my digestion. I used to rely on an electric

wheelchair occasionally. I had it for several years but it never sat well with me (excuse the pun). Then one day I decided to return it and give myself no choice but to focus on strengthening exercises. I pushed myself to remain on my feet and I have managed to do just that. I found my 'sea legs' on land.

All those years ago, my doctor was not certain about my diagnosis and suspected it might be something else. Three years ago, based on my symptoms and the progression of the illness, I was diagnosed with Hereditary Spastic Paraplegia, (HSP). My diagnosis is a combination of SMA and HSP, another balancing act in my repertoire! There is not much difference between the symptoms of these two diseases but the spasticity that I present makes my life much more challenging than if it were SMA alone.

Despite my health issues, I completed my studies and had a job as a community worker for different organizations. Now, because of my condition, I volunteer a few hours per week. This keeps me busy, and it is rewarding for me to be there for others. I will not hide that some days it is very difficult to confront the routine with all the physical pain and weakness that I feel. What keeps me going is my deep trust and love for God. With faith, I am dealing with my condition with gratitude and grace.

I avoid taking prescription medication because of unexpected side effects that can be worse for me than the average person. I do take vitamins, especially B complex, D, B2, magnesium, and collagen powder to help relieve physical pain and headaches. Additionally, I do chair yoga, Aquafit, and stretching exercises based on videos online. Exercise keeps at bay a lot of spasticity, tiredness and the bad mood tied to bodily discomfort.

I am well supported by family and friends. Their unconditional love and support are truly a blessing. I also have a great team of professionals with whom I consult often. I try my best to stay positive and hopeful in my life. I attend events that are inspiring and educational, keep encouraging quotes in mind, read books, and listen to motivational speakers. All of this teaches me about myself and how to be at ease with my situation. Life has taught me about finding equilibrium while staying in motion. This wheel will keep on spinning as long as it can.

Learning to Overcome the Challenge of HSP

By Kyle Alumbaugh, HSP

I was born in 1979 in Marion, Indiana. Hereditary Spastic Paraplegia runs in our family; my dad Jack and brother Ben also have HSP. Dad raised us by himself while we were both very young. He worked in several factories, and by his early thirties, HSP had impacted him enough to make it difficult to work. He understood firsthand the effect of spastic paraplegia and made sure that my brother and I enjoyed many of the same things as other kids. Playing sports, running, riding bikes, and other things were a bit challenging, especially since my left foot was at a 90-degree angle, but I didn't allow these challenges to stop me.

Growing up, some kids thought Ben and I were different and tried to make life miserable. I remember an instance when I was 12 years old. As the school bell rang, a kid named Shawn said, "Kyle, you're gonna be late for class!" I knew he was setting me up to trip me as I rushed past. I managed to jump over his leg and whack him as I went by. I learned early that many times with challenges just as this one, we must stand up to the task and take action.

In eighth grade, I was scheduled to have a tendon lengthening surgery with doctors in Rochester, Minnesota. This allowed my left foot to be flat. My right foot wasn't as bad. The doctors ended up putting pins in my left leg, and a cast on my right leg. After some healing and therapy, I was able to walk much better. However, I was told that due to HSP being progressive, I would still have difficulty as I aged. I knew this, but I was going to take it one a day at a time and do my best. During this time I was also diagnosed with epilepsy. I'm not sure if epilepsy is part of HSP, being a neurological disorder, but it was a tough challenge for many years. Since 2018, I have been epilepsy free.

In my twenties, I started using a cane. Canes can actually give a hip, cool look. Some days, I'd put on my fedora and head out the door. By my late twenties-early thirties, I would sometimes use a walker instead of the cane.

In 2000, Ben had a wonderful daughter named Meghan. She may have some elements of HSP,



Kyle (left) with his dad Jack (center) and brother Ben

but not nearly as severe physically as her dad, her grandfather or me. She's currently doing well as a manager in the fast food industry.

In November 2002, I married Gloria. Eventually, we moved to southern Indiana where she was born and raised. There I worked in retail while going to college for a degree in business. A key thing that helped me was Vocational Rehabilitation Services (VRS), a state-funded service. (It's worth checking to see if your state has similar programs.) VRS provided funding for wear and tear on my vehicle going to and from college. In addition, they made my house and car wheelchair accessible. This was crucial since, midway through college, I was falling a lot and decided that a wheelchair was needed.

I finished college in 2016 and received my bachelor's degree. Getting a job was tough. Being in a wheelchair does have its challenges in many job areas. I am currently working part-time as a financial advisor at a bank.

What can I say about HSP? Having any disease, whether it is spastic paraplegia, cancer, Parkinson's disease, epilepsy or any other, doesn't make life easier. So what do we do? Although it is a challenge, we must learn to take joy in it and find a solution to overcome it.

When we think of what the future looks like in comparison with our current position, it can cause us to create assumptions, have fear and worry, and cause us to get down on ourselves. For me, I find joy and relief in Jesus Christ. I find that the many challenges of life have solutions in His word. In addition, I have a loving and supportive wife. My advice is be strong, make good choices, and find ways that help you pull through and overcome this challenge of HSP.

Ten Steps for Coping with Chronic Illness

Submitted by Mary Ann Inman, PLS

Note: This column was modified from Health News from Harvard Medical School



Mary Ann Inman

1 Seek information from reliable sources.

Learning about your condition will help you understand what's happening and why. First, direct your questions to your medical care providers. If you want to do more in-depth research, ask them about trusted sources of

medical information. For example, some clinics and hospitals excel in neurological care and research. You can also visit sp-foundation.org for brochures about HSP and PLS that you can print.

2 Expect your doctors to become your partners in care.

Don't leave everything to your doctor; take responsibility for your care. Listen to your body and write down changes. Learn to check your blood pressure, weigh yourself, and log your symptoms. Self-advocacy helps you spot potentially harmful changes before they bloom into real trouble.

3 Build a team.

Seek out other experts in addition to your doctor. Physical, occupational, and/or speech therapists are great resources. Dietitians can help you with nutrition information.

4 Don't assume HSP or PLS is responsible for all symptoms.

It's sometimes easy to chalk up your health issues as part of your HSP or PLS. Make sure that the "whole you" receives the appropriate care, diagnostic testing and treatment. Having an excellent and responsive primary care physician to guide your specialty care is a good start to creating the full picture of your health.

5 Make a healthy investment in yourself.

Treatment for almost any chronic condition involves lifestyle changes: stop smoking, lose weight, exercise more, and eat healthy. Although these steps are sometimes put on the back burner, they are important. People

who are motivated to make such changes are more likely to successfully manage a chronic condition. Investing the time and energy to make healthy changes usually pays off, ranging from feeling better to living longer.

6 Make it a family affair.

The lifestyle changes you make to ease your chronic conditions are good for everyone. Instead of going it alone, ask family members or friends help you.

7 Manage your medications.

Know about the drugs you take — why you take them, how best to take them, how they potentially interact, and side effects. Talking with your medical provider or your pharmacist can put drug information into perspective.

8 Beware of depression.

One third or more of people with chronic diseases are affected. Depression can keep you from taking important medications, seeing your doctor when you need to, or pursuing healthy habits. Let your doctor know if you think you're depressed or heading in that direction. A specialist can help you mentally manage setbacks and progression.

9 Seek support from groups.

Doctors, nurses, and other health care professionals don't necessarily know what it's like to live with afflictions daily. Look for a support group in your area and talk with people who have been through what you are facing. [I highly recommend joining the Spastic Paraplegic Facebook page and attending face-to-face or online SPF Connection events. - Mary Ann Inman]

10 Plan for your care long-term.

Install grab bars, ramps, and obtain mobility devices before you feel you need them daily. While HSP and PLS are not fatal, they can be debilitating. A proactive approach toward planning for your future needs can help make life smoother. Also, consider having a Power of Attorney and Living Will to make your health care wishes known. Spelling out, in writing, the care that you want until the very end can save you and your loved ones frustration, confusion and anguish.

Grab a Chair and Sit to be Fit

Sometimes it's tough to be motivated to exercise. Daily demands, packed calendars and hectic family schedules can be exhausting. Couple that with mobility, stability or balance issues and it can be easy to opt for sitting it out. No worries! Grab a seat for a chair-based workout. Chair exercise routines can be designed for all levels of ability and bring multiple benefits, such as:

- **Improved strength and muscle tone,**
- **Better coordination, balance and flexibility,**
- **Greater range of motion,**
- **Improved circulation, and**
- **Heightened confidence.**

When creating a chair-based exercise program, there are several factors to consider.

How to Start.

Begin by getting medical clearance. Talk with your doctor, physical therapist, or other health care provider about activities suitable for your medical condition and mobility issues. Your doctor or physical therapist can help you find a suitable exercise routine. Key questions to ask include:

- **How much exercise can I do each day and each week?**
- **What type of exercise should I do?**
- **What exercises or activities should I avoid?**

What You Need.

First, find a sturdy chair that's wide enough to allow you to move freely. Don't use one with wheels. The chair should be at a height that puts your hips at or above knee level.

Next, choose the equipment that's right for you. Equipment like free weights, ankle weights, exercise bands, yoga blocks, stress balls, and under-desk bikes can be incorporated into a chair workout.

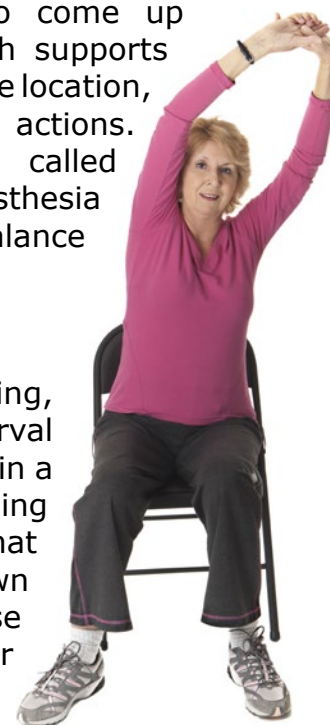
Put on some inspiring music and comfortable clothes that allow you to move easily and you're ready to go.

How to Position Yourself.

Sit toward the front of the chair, place your feet firmly on the ground, and press your weight into them. Positioning yourself correctly allows sensory information to come up through the feet, which supports the body's ability to sense location, movements, and actions. This sensory input, called proprioception or kinesthesia provides a sense of balance and spatial orientation.

What You Can Do.

Yoga, dance moves, boxing, karate, tai chi, even interval training all can be done in a chair. The important thing is to find activities that you enjoy, go at your own pace, and make exercise an essential part of your life.



Online Chair Exercise Resources.

There is a wealth of information, videos and classes to explore online. Some sites to get you started are:

Zumba exercises with Nancy D.

https://www.abovems.com/en_us/home/ms-wellness/exercise-fitness/zumba-exercises.html

Stationary cardio with Nancy D.

https://www.abovems.com/en_us/home/ms-wellness/exercise-fitness/stationary-cardio.html

Silver Sneakers Total-Body Chair Workout (Available at no cost for adults 65+ through select Medicare plans; check for eligibility): silversneakers.com

Sources: www.hspersunite.org.au; Brain & Life Exercise, December 2020/January 2021; www.helpguide.org/articles/healthy-living/chair-exercises-and-limited-mobility-fitness

You Asked, We Answered: FAQs About PLS

By Pam Lanter, PT, DPT, NCS, University Hospitals ALS Care Committee



Pam Lanter, PT, DPT, NCS

The following Q&A was compiled to provide insight into some of the most frequently asked questions about PLS.

HOW MANY KNOWN CASES OF PLS ARE THERE IN THE US? THE WORLD?

Due to the difficulty in diagnosing PLS and distinguishing it from other disease processes, it is tough to determine a definitive number. It is estimated that about 500 people are currently living in the U.S. with PLS, although some researchers believe that the number might be 1,000-2,000. In a *Lancet* article from 2018, 330,918 people worldwide were identified as having been diagnosed with a MND which included PLS, HSP, amyotrophic lateral sclerosis (ALS), spinal muscular atrophy (SMA), progressive muscular atrophy (PMA), or pseudobulbar palsy.

IS THERE A SPECIFIC GENE/GENES ASSOCIATED WITH PLS?

The underlying cause of adult PLS is currently unknown. In most cases, it occurs sporadically in people with no family history of the condition which makes it difficult to find a genetic component. In 2008, an article was published in the scientific journal *Archives of Neurology*, (now known as *JAMA Neurology*) about a large family that had eight individuals diagnosed with adult-onset PLS. Those researchers found one common spot on a specific chromosome (4ptel-4p16.1) in the affected individuals. In contrast, HSP has been associated with over 70 genes so there appears to be much more of a genetic component of HSP than PLS. Multiple mutations of the ALS2 gene have been identified that cause juvenile PLS.

WHAT IS THE PLSFRS AND HOW DOES IT DIFFER FROM ALSFRS?

The Primary Lateral Sclerosis Functional Rating Scale (PLSFRS) is a tool that physicians use to measure the progress of PLS. It has been studied by researchers and found to be better at detecting the slower changes associated with PLS than the Amyotrophic Lateral Sclerosis Functional Rating Scale - Revised (ALSFRS-R). When comparing the two scales, the PLSFRS has two levels of intermediate function added to the first 10 items

of the ALSFRS-R which increased the maximum score from 48 points to 68 points. Generally, the higher the score on the PLSFRS, the less symptoms a person has to manage.

HOW IS THE DIAPHRAGM AFFECTED IN PLS?

The short answer is that patients with PLS may end up having muscle weakness in their diaphragm which could lead to some respiratory difficulties. For more details, we need to look at how people breathe. The act of breathing is controlled by lower motor neurons and usually is involuntary, so we don't have to think about it to continue to breathe. The diaphragm is a muscle controlled by the phrenic nerve that contracts and flattens as you inhale and relaxes as you exhale. The phrenic nerve (controlled by lower motor neurons) is rarely affected in PLS and helps explain why there is minimal evidence of respiratory compromise.

IS THERE ANY DIFFERENCE BETWEEN BULBAR ONSET AND LIMB ONSET AS FAR AS PROGRESSION OR SYMPTOMS?

The main difference between limb onset PLS and bulbar onset PLS is patients with limb onset tend to have symptoms earlier in life than patients who initially have bulbar symptoms. There does not appear to be much difference in how quickly the disease progresses. Ninety percent of people with PLS start noticing symptoms in their legs (limb onset) which makes it the most common form of PLS. Some people start noticing symptoms in one side of their body (e.g., right arm and right leg) or in both arms, though this is much less common. People often first report stiffness or clumsiness/balance problems when they have limb onset PLS. In bulbar onset PLS, patients tend to initially experience difficulties talking, especially the rate at which they speak. They can also report difficulties swallowing.

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7. Raquel Gil Gouveia, Anabela Pinto, Teresinha Evangelista, António Atalaia, Isabel Conceição & Mamede de Carvalho (2006) Evidence for central abnormality in respiratory control in primary lateral sclerosis, *Amyotrophic Lateral Sclerosis*, 7:1, 57-60, DOI: 10.1080/14660820510012031

Take the Challenge During HSP and PLS Awareness Week August 22-28, 2021

The 2021 Awareness Week theme is “Challenge.” The goal is to inspire global awareness and actively promote the message of rare disease medical research for #HSPandPLS.

During Awareness Week join us each day at 1:00 p.m. Central Time for a global Zoom conversation with new and old HSP and PLS friends.

Sunday, August 22 is Sports Day

Wear your favorite sports team apparel; record yourself playing a favorite sport or cheering on your favorite team; tell us about your plans to host or participate in a run-, walk-, or roll-a-thon or any other fundraiser for #HSPandPLS.

Monday, August 23 is Hobbies Day

Tell about your favorite hobby or something on your “wanna do” list.

Tuesday, August 24 is Sunglasses Day

Strike a pose in your favorite pair as we shine a light on HSP and PLS.

Wednesday, August 25 is Creative Arts Day

Get creative and share your talent. Music, painting, acting, photography, writing, dance or any other creative endeavor is welcome!

Thursday, August 26 is Swag Day

Wear your favorite SPF items and get stylin’. Don’t have any yet? Go to the online SPF Store via the sp-foundation.org homepage; select “SPF STORE” and click on the yellow “SHOP” icon to find your new favs!

Friday, August 27 is Aid Day

Share your insight into how using a cane, walker, wheels, functional electrical stimulation (FES) or other mobility aid helps keep you on the move.

Saturday, August 28 is Celebration Day

Join us for a special day of presentations and recognition.

REGISTER FOR FREE

at sp-foundation.org.

Look for the Awareness Week link.



Look Good. Do Good. Shop the New Online SPF Store.

Fun, functional and fashionable items are just a click away with the Spastic Paraplegia Foundation’s online SPF Store. Items from the official site for branded SPF items help raise awareness of HSP and PLS. Additionally, a portion of each purchase benefits medical research through SPF.

Show Your SPF Style

There are so many different items being added all the time, including:

- T-shirts, hoodies and other logo wear for men, women and children
- Tech Accessories such as Phone and Tablet Cases
- Tote Bags and Backpacks
- Mugs for Home, Office and Travel
- Stationery and Stickers
- Plus so Much More

See all of the products, designs and colors by visiting sp-foundation.org/SPF-STORE and then clicking on the yellow “SHOP” icon.



Tina's Tips: My NuStep

By Tina Croghan, HSP SPG7, SPF Board Member

After I broke my ankle in October 2020, I was lucky enough to be able to do intensive rehab at SSM Lake Saint Louis Day Institute and Rehabilitation. While there, I had Physical Therapy (PT) and Occupational Therapy (OT) three times a week. One of the many tools PT used to help me increase my leg strength was a recumbent stair step machine made by NuStep.

Since I could no longer perform the rotational movement required on my 10-year-old recumbent bike, I purchased my own NuStep machine, the T4R model. Of course the one I used in PT was the top-of-the-line commercial grade with all of the bells and whistles; I didn't need all of those features at home.

This equipment doesn't change the fact that I have a degenerative, upper-motor neuron disease that requires me to do many kinds of exercises and stretches to stay strong and have good muscle tone. Having a NuStep just makes exercising more efficient and fun for me!



Tina "climbing stairs" with her NuStep.

JOHN STAEHLE 1946-2020



SPF board member, *Synapse Newsletter* Senior Editor 2013 to 2020, North Texas SPF Ambassador 2005 to 2020.

THE STAEHLE LEGACY LEAGUE

Members of The Staehle Legacy League help to assure a future HSP/PLS cure by naming SPF in their will, trust, retirement plan, life insurance policy or annuity.

For more information about planned giving, please contact us at JohnStaehleSPFLegacyLeague@gmail.com or call (877) 773-4483

THE STAEHLE LEGACY LEAGUE MEMBERS

Frank Davis

Linda & Craig Gentner

Britt Gibson

Dave Handley & Pamela Jordan-Handley

Dina & Chris Landphair

GOOD NEWS!! THE SPF 2020 ANNUAL REPORT IS NOW ONLINE!

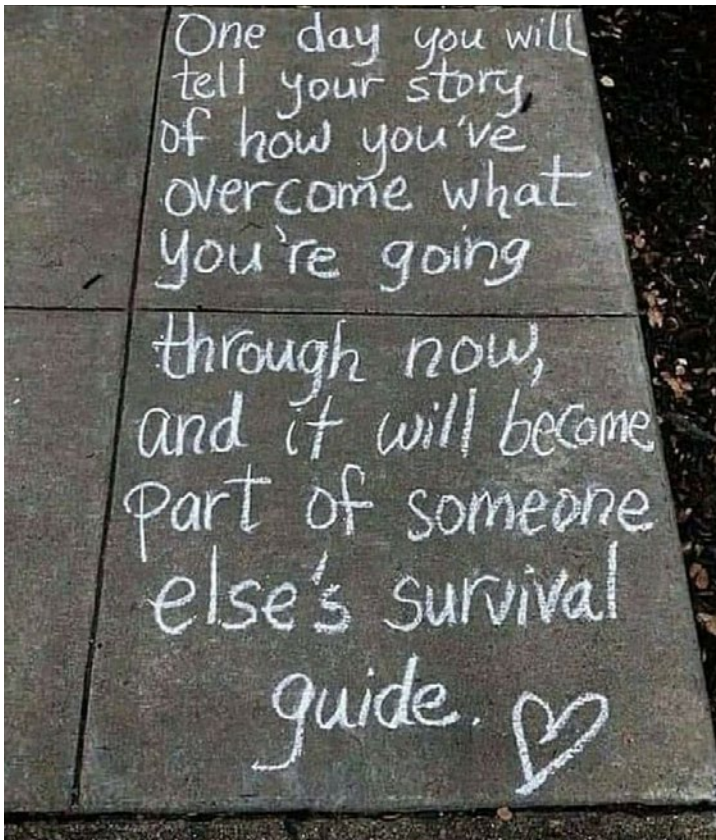
Just enter: https://bit.ly/SPF_AR

Dr Fink's Neurological Support Group



Join the Official SPF Group at:

<https://www.facebook.com/groups/HSPandPLS/>



Everyone has a Story to Tell. What's YOURS?

Work on the Fall edition of the *Synapse* is underway and we want to share your story!

Synapse, the Spastic Paraplegia Foundation's quarterly publication, reaches people across the country and around the globe. Readers are eager to hear from others with Hereditary Spastic Paraplegia (HSP) and Primary Lateral Sclerosis (PLS).

Your story can let them know they are not alone in their quest for a diagnosis. Your knowledge and insight gained along the way can help others thrive. Your articles, poems and musings can brighten their day.

Articles can be any text format; photos can be any picture format.

Here are some topics to spark your creativity:

- Ways you keep doing what you love — modifications you make to keep active and stay in the game.
- Technologies and devices that can help make life easier.
- Tips for stretching, exercising, balancing and staying as physically active as possible.
- Milestones celebrated on your journey.

- Sharing a different perspective — how caregivers, partners or children experience and support their loved ones with HSP or PLS.
- How you keep your sense of humor and view the absurdities of life.
- Fundraisers, Connection get-togethers and other events you do to promote awareness and support the Spastic Paraplegia Foundation's mission of finding a cure.
- Proving that a picture is worth a thousand words — share your photos from HSP and PLS Awareness Week or other special events.

You don't have to be a writer to submit an article! You just need a passion for sharing what keeps you going and growing while living with PLS or HSP.

To submit an article or discuss your ideas contact synapsenewsletter.spf@gmail.com

We look forward to hearing from you!



SPF VISION

The day when all individuals with HSP or PLS are diagnosed, treated and cured.

WHY YOUR DONATIONS ARE IMPORTANT

Donations to the Spastic Paraplegia Foundation are crucial to funding research to find a cure for HSP and PLS. We ask for donations because the more dollars we receive, the more leading research grants we can fund. Your donation allows us to support the work to find a cure.

We do not have an office. We do not have employees; we are all volunteers - from the President and SPF Board of Directors to the Scientific Advisory Board members and State Ambassadors. Last year 92% of donations went to research. Visit our website to see the many ways you can donate, fundraise and be involved.

1605 Goularte Place
Fremont, CA 94539-7241



It is Essential to Register with the Spastic Paraplegia Foundation!

Medical researchers contact SPF to locate people with a specific gene mutation. Upon request, we provide a list indexing people with a specific gene mutation from the SPF database. If you are registered, then you may be selected for medical research or gene therapies.

Join at SP-Foundation.org/news-resources/stay-informed.html

