



HSP Newslink

The Newsletter of the Hereditary Spastic Paraplegia Support Group
Registered Charity No. 1109398

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HSP & FSP – Hereditary Spastic Paraplegia,
Familial Spastic Paraplegia & Strumpell-Lorrain Syndrome

Christmas arrived early for Steven Foot



The priceless moment Steven was stunned when he realised he was being presented with a TGA Breeze

Despite living with Hereditary Spastic Paraplegia, Steven Foot from Marnhull, Dorset, has been able to regain independence outdoors and re-live his passion for the countryside and point-to-point horseracing, thanks to a community-donated TGA Breeze mobility scooter with off-road capabilities. Before his condition became too severe, Steven owned a successful equestrian transportation business that fulfilled his love of the outdoors.

Following the reluctant sale of his business, Steven was forced to spend considerably more time indoors with restrictions on his outdoor independence – this has just changed thanks to the support of his local community. Instigated by the HSP Support Group and its relationship with The Old Thatch pub near Wimborne, donations have been raised to purchase a new, state-of-the-art TGA mobility scooter so Steven can once again access the countryside.



Steven Foot on his new TGA Breeze S4 GT mobility scooter with Ian Bennett, the girls who jumped out of an aeroplane and the landlady of The Old Thatch

Customers and staff at The Old Thatch pub have worked together to raise funds to purchase the new scooter. This is the same type of scooter that won the Top Gear extreme mobility scooter challenge, which was broadcast on BBC2.

Steven will now be able to tackle local rural tracks and fields on his TGA scooter as it has an 8mph top speed, wide high-grip tyres and extra power and performance. Owning this Breeze has only been made possible by several fundraising activities since July. Donations have been gathered from a sponsored skydive by four members of pub staff and relatives, collection tins, live music, BBQ's and car washing at The Old Thatch. These funds were dramatically increased when, in support of a local resident in need, the Sturminster Newton Round Table made a generous donation.

The Old Thatch was keen to support the local community so it was agreed that we'd raise funds to help a Dorset resident affected by HSP. Following a period of communication with all Dorset members of the HSP group, Steven was selected as an ideal recipient. We were all determined to pull together to raise funds so Steven could regain freedom again outdoors. I would like to thank everyone so much for helping us reach this target especially my great friends the Roberts family, Pete, Sarah, Gemma and Tanya. Thanks also to the crazy female skydivers (three of whom are members of the Roberts family), the management staff and customers of the Old

Thatch, Sturminster Newton Round Table and TGA who gave us a discount on the Breeze. TGA were moved by our story and have been so supportive in terms of scooter supply and delivery. When we organised the surprise party and presentation at the pub for Steven, his reaction when he appreciated what was going on was priceless! As can be clearly seen on the headline picture he was so overwhelmed and emotional. What a special occasion it was, made even better with a local band called Wayward, entertaining us all for free in support of Steven. This scooter will change his life no end as Steven is now free to drive around the countryside and is already planning to attend point-to-point events once more.

Steven added: "I am overwhelmed by the generosity of my local community and friends and can not thank everyone enough."

We also used some of the funds raised to purchase a portable ramp for a member who lives near Poole. Being a wheelchair user, it enabled the recipient to access his son and daughters houses with ease.

The Old Thatch is now planning similar fundraising activities to support local individuals affected with HSP. Now we have targeted Dorset, I will be shortly writing to all members within 50 miles of the pub to see if there is any mobility equipment or aids that may be of use. I'll then hopefully hear back from one or two of you, giving all at The Old Thatch a target to knuckle down and aim for.



Steven outside his local pub on his new TGA scooter

Chairman's Column

Although a month has now passed by, I hope you enjoyed the festivities and wish you all the best for 2015.

You will notice from the articles on the front of this and the previous Newsletter that I have been involved with some amazing work at my local pub. With the help and support of many lovely people it has been great fun and presenting a scooter to Steven Foot, who knew almost nothing about it, was one of the highlights of all the charity work I've ever been involved with. It was a very emotional evening which won't be forgotten. With the help of TGA Mobility who supplied the scooter, we are getting plenty of media coverage out of the whole fundraising experience which is all helping raise awareness of HSP.

I know many other people are involved in fundraising for us and please keep up the great work. I know of four individuals who are preparing to compete in half marathons and marathons for us within the next two or three months including the London Marathon. The ever reliable Peter Bateman continues to bring in the funds from his collection boxes scattered around the Porthcawl area of South Wales. Many thanks to everyone who fundraises for our cause, your efforts are much appreciated and put to very good use.

Since the last newsletter I have become involved with Riding for the Disabled, and every Friday morning I'm now in the saddle for at least 20 minutes. I used to ride so it's good to get back into it. Although it's a little frustrating how useless my legs have become whilst in the saddle, it's great physiotherapy and I've met some lovely people. The lady who runs it is called Dianne Tubbs and she won a silver medal in the Atlanta Para-Olympics for dressage.

In October, my elder daughter was 21 and a month later my younger daughter was 18. Part of the celebrations consisted of a family trip to Majorca. Although we only went for four days, we had a fantastic time.

It was the first time I've taken my wheelchair abroad and it made the whole experience far more enjoyable. There were no problems at the airports and my family were able to push me from hotel to beach to cocktail bar, in the lovely October Mediterranean sunshine. It made all the difference and when I returned home, instead of being totally wiped out, I felt like I'd had a relaxing holiday. I certainly won't travel abroad again without taking my wheelchair.

I've been making the most of my season ticket for Bournemouth football club. This is something else I do in a wheelchair and my carer (who is one of my closest friends) is admitted free of charge. It's not just a football match, it's a full day out and we've made many new friends. I'd recommend it to anyone, even if you're not the greatest football fan.

I've managed to maintain my flying with Aerobility although the recent weather hasn't helped. I'm currently training to fly their new aircraft which is very economical and consequently much cheaper to hire.

My garden also keeps me busy even at this time of year. I try to limit myself to a maximum of 30 minutes work a day or I find fatigue sets in for a couple of days and prevents me doing anything. This year I've set myself a fun challenge of having something in flower every day of the year. I've just noticed that snowdrops are coming into flower and thanks to a very early flowering Camellia that has been in bloom since New Years day, I'm confident that I'll meet the challenge. I love my garden and without the use of a four wheeled rollator, I couldn't do half the jobs I do outdoors.

You can see that although the old legs are getting wobblier and I'm definitely slowing down a little, I always try to keep myself busy. I think one of the secrets is not to be afraid to use help when required. I still love my new mobility scooter and I'm now making far more use of my wheelchair.

Partly because of my busy lifestyle but probably more on account of my

progressing HSP and the knock on effects, I feel the time has come to begin lowering my workload somewhat. I'd like to begin this process by standing down as Region 4 Co-ordinator. I did ask for a volunteer to take on the role at a recent Ashburton meeting and had no takers, but not everyone was in attendance. If you are interested in becoming Region 4 co-ordinator, please let me or any committee member know. Those who have attended Ashburton or indeed any other regional meetings will appreciate how important these get-togethers are.

I feel it's time we had new blood on the committee, but please only consider this if you have some time available. One role I'm happy to continue is editing the newsletter but this is of course much easier when I receive material from members to be included. If I'm short of material this will only cause a delay because I'm not happy issuing newsletters unless they are an interesting read. Many thanks to those of you who have contributed to this issue and as always I'd like to thank Adam Lawrence for his column.

I'd like to thank Simon Hubbard for all the work he does on our behalf communicating with the medical profession.

Further thanks to John Mason who has constructed a new web linked database that will improve how we manage and communicate with our membership. Area co-ordinators can be given access to this new facility so that they can keep totally up to date with the members in their regions.

You will notice that more local meetings are now taking place around the country. These meetings are very important, giving members the chance to meet others with HSP, share information and no longer have the feeling that many of us know of being 'alone'. Thank you to Irena Pritchard, Barbara Jones, Penny Cohen, June Masding and Hilary Croydon for all your hard work in setting up regional get-togethers in St Helens, Norwich and now also Birmingham and Colchester.

I mentioned in the previous edition that our Group had some money to give away to researchers. Back in September the committee got together to discuss the applications we'd received and I am happy to tell you that after much discussion we decided we were in a position to support the following four research organisations.

- Plymouth University - £2000 for final phase of research project
- University of Cambridge - £8000 for 1st year of project
- National Hospital - £9,700 for Genome Registry set up
- Sheffield Project – We have agreed to offer a maximum of £15,000 to this work as costs and timescales become clearer

It is fantastic that we are able to help the medical experts and hopefully one day we'll be in a much stronger financial position enabling us to provide researchers with much larger grants.



Sue Bailey (Right) presenting a cheque to Kate Winstone from Plymouth University during a meeting at Ashburton

Our Facebook group, Hereditary Spastic Paraplegia's Unite now has in excess of 550 members from all around the globe. There are communications taking place on a daily basis so if you haven't yet signed up, please do so.

I look forward to meeting up with members at forthcoming events and I'm always on the end of the phone if required.

Ian Bennett

Research Column

In the September 2014 Newslink I compared HSP with other types of Motor Neuron Disease, in which I summarised life expectancy for people with HSP as "The majority of individuals with HSP have a normal life expectancy." I had 'borrowed' the text for the description from other HSP sites. I was asked by one reader about this aspect which is an interesting question, and potentially a sensitive issue. My advice for any readers who are concerned about this should contact their HSP specialist to get their own answer.

Some clarity is obtained from the NINDS site (http://www.ninds.nih.gov/disorders/hereditary_spastic_paraplegia/hereditary_spastic_paraplegia.htm) which indicates that "The majority of individuals with uncomplicated HSP have a normal life expectancy." I realise that I should have included this important differentiation in my column. There is an increased risk with uncomplicated/pure HSP due to falls and from infections, principally bladder. Given that life expectancy for uncomplicated HSP is usually normal this raises the question about what is the life expectancy for complex HSP. In summary, there is not much information which I can find, and this is an overview of what I have found.

In my review of research papers there are only two papers which talk about life expectancy, both in relation to SPG11. There is a Dutch study covering 18 patients (<http://www.ncbi.nlm.nih.gov/pubmed/23443022>) and a study from France with 7 patients (<http://www.ncbi.nlm.nih.gov/pubmed/21035867>). The Dutch study suggests a life expectancy of 3-4 decades after the start of walking problems. The paper from France gives life expectancy of 30-40 years, but it is not clear from the abstract if this is from onset, the same as the Dutch study, or 30-40 years total. I note that the number of patients in these studies seems quite small, which makes me question the confidence.

Perhaps life expectancy actually varies significantly from person to person, and perhaps it can be varied with the level of care that is received.

There is also a lot of detailed information about HSP on this Washington University page (<http://neuromuscular.wustl.edu/spinal/fsp.html>) which indicates that SPG11 has an onset usually before 20 but sometimes up to 50, which perhaps ties in with the Dutch study in that some people don't get SPG11 until older than 30-40. The Washington page doesn't give life expectancy for SPG11. It is not clear where all the data comes from on the Washington page, whilst there are references in most areas of the page, I don't have access to full papers to check. This Washington page indicates that SPG13 has a life expectancy of less than 2 decades after onset.

Looking for further information starts to become more of a challenge. All of the other information/comments I find are not specific and I can find various answers to the question which contradict each other. Since I don't know which to believe, if any, I'll avoid any of those links here.

Meanwhile, I've downloaded the results of my survey and have started the analysis. In the end I've had around 90 responses, predominantly from the UK and US. I have respondents with a fair range of mobility's and there seems to be a wide range in the number of medications being taken. Stretches seems to be the most frequent form of exercise/relaxation. I've quite a bit of analysis to do before the end of February!

Adam Lawrence

HSP Group Funds

Funds are available for members to apply for financial assistance with the purchase of mobility aids or equipment that may improve quality of life or simply make life easier. If interested, please request a grant application form from a committee member.

Useful Information

The Americans have created a useful, informative HSP booklet on their website which can be downloaded. The link to access this is:

<https://sp-foundation.org/hsp-booklet/>

Members' Letters

The Duke of Edinburgh's Award

I expect you are all familiar with the Award, as we were when Tim was growing up, but we didn't realise how much it caters for people with disabilities as well as for those who can tramp for miles over rough country!

For anyone who hasn't come across it before, the Award seeks to provide a framework within which young people between the ages of 14 and 25 can challenge themselves in a range of areas. There are three levels of the Award – Bronze (14+), Silver (15+) and Gold (16+) – and for each it is necessary to spend a few months pursuing a sport, a skill and service to the community. In addition, the participants do an expedition in a team, involving travelling on land or water and camping. At each level the times increase. For Gold there is an additional element of a residential stay of four days away from home with people you don't know.

Tim did the Bronze Award when he was still at school before the physical onset of HSP. Then he set about doing the Silver Award while he was at college. For this the team had to camp for two nights and walk for several hours each day. On the second day of walking Tim's left hip suddenly went weak but he was determined not to give up so he carried on walking despite pain in his hip. On the third day the D of E assessors were concerned about Tim and decided not to allow the team to do any more walking. Instead, they would spend more time on the project element of the expedition, producing an extended presentation about the area in which they had walked. Tim

went to the doctor and, two years later, had a genetic diagnosis of one of the complex forms of HSP - SPG11.

Tim has gained enormous satisfaction from doing the Award as well as having had a lot of fun and making new friends.

He was very keen to do the Gold Award as well and so we talked to D of E to see what could be done. It turned out that there is considerable flexibility within the scheme to enable people with disabilities to take part. For example, the expedition can be done in a wheelchair (powered if necessary) or a canoe and there are specialist providers approved by D of E who can organise an expedition to suit differing people's needs. As long as it is a challenge for the participants and there is a minimum number of four people to ensure the element of teamwork there can be quite a lot of variation from the standard expedition.



The specialist providers can also provide the residential element for the Gold Award and Tim did this last August with The Bendrigg Trust on the edge of the Lake District (see photo). He loved it – not a parent in sight for four days! It is perfectly possible to take a carer, if necessary, but Tim was quite happy to have a bath when he got back!

We have been actively in touch with The Bendrigg Trust for two years trying to arrange an expedition but they have been

struggling to find the minimum number of four people to do an expedition at the same time. As the Award has to be completed before your 25th birthday and Tim will be 25 in July 2017 he really needs to do the expedition in 2015 or 2016. Tim has now done every aspect of the Gold Award except for the expedition and he is becoming very worried that he will be unable to finish the Award solely because enough people are not available.

So he is wondering whether you, or any of your friends, know of anyone, with a disability or able bodied, between the ages of 16 and 25, who would be interested? It is not necessary to have a disability to go through a specialist provider. It simply means that the expedition will not be quite the same as the usual trekking for miles with a heavy rucksack for several days. The challenge is a little different. There is more emphasis on the project element and less on the walking. Although there is the necessary travelling it may be a mix of walking/wheelchair and canoeing, for example. Sometimes, the group may not camp but travel out from a base each day. The expedition is designed to be challenging for everyone.

So, for those who have already done the Bronze and Silver expeditions a Gold expedition with a difference might make a refreshing change.

However, it is not necessary to have done either the Bronze or Silver award first. It is possible to go straight in at Gold and the expedition could be the first part of the award to be achieved.

The Bendrigg Trust's website is www.bendrigg.org.uk. Their phone number is 01539 723766 and the two people primarily dealing with D of E are Jenny Jackson and Jo Winchester.

The D of E website is www.dofe.org. Do have a look.

Our phone number is 01284 728242 if you would like to know more. We would love to hear from you.

Hilary Croydon

The Future is Bright!

I was diagnosed with HSP three years ago and it was quite a blow. In some senses though it was also a huge relief because I had been tripping and falling for some time and I realised something was not right. The diagnosis was at least an explanation. I was a broadcast journalist and would always trip on a tiny step on the studio floor.

There are awful disadvantages to being disabled in the modern world but there are also enormous advantages. New technology is one of them. I now edit an investigative website called 'Wales Eye'. All of this I can do sitting down using my computer. My computer has become my window to the world. I research stories on the internet, talk to correspondents, write material which I then publish, all from one place. It is brilliant!

In the old days as a journalist you would always have to be moving around, finding out information and delivering your script. But not any more. The internet, if used wisely, can be a huge resource for disabled people. Yes the information is unfiltered and can be disturbing.

After my diagnosis I made the fatal mistake of going on the internet to find out about HSP and, of course, read all the worst scenarios. The other stories of people continuing to live long and fulfilling lives were somehow ignored by my frantic brain. But on the other hand all the information is out there, and if you know how and where to look it is a huge benefit which gives those diagnosed with the condition an enormous advantage when it comes to talking to professionals about treatment. Once you learn which websites are reputable and can be trusted you are away. The information from – for example – government ministers is all posted on the internet and departments usually have their own websites.

Email too is a tremendous tool. Almost all media outlets for organisations have an email address and as long as you can strike

the keyboard characters, you can gain information. You can create your own website, and publish items, again all while sitting in the comfort of your own home.

As a side issue deadlines too are a thing of the past. As a cub reporter on a local evening paper, the deadline times were drummed into my head – 9.45 am for inside pages, 10.45 am for the front page. Now as soon as the story is ready it can be published. Again this has its critics who say there is no time for reflection and critical mistakes are made. This can be true, but it also means you can reach a wide audience more or less instantly so once more, if new technology is used wisely it is a tremendous boon.

HSP is incredibly slow, and does at least give you a chance to adjust. The main problem for me is less the physical aspect than the mental – knowing it can only ever get worse.

But new technology does at least help.

Phil Parry Region 6

Regional News

Region 5

East Anglia Colchester Meeting

We held our first meeting in this part of the region on Sunday, 21st September at the Holiday Inn, Eight Ash Green, near Colchester.

Altogether, twenty three of us attended including one or two potential new members who we were delighted to see. We had an informal get together, meeting new people and catching up with old friends. It was lovely to see Della and Roger, and Mo and Lorraine who had joined us from outside the region. Thanks to them for coming along. Members from all regions are very welcome.

We had a chat about the way forward and the consensus was that the members present would like to meet up in a village hall type

venue with plenty of space to make it easy to move around and talk to different people. It was felt that although there would be the cost of the hall to consider we would be able to arrange our own refreshments which would be cheaper and more informal. It will also give us scope to arrange speakers in the future, if that is what members would like.

We are in the process of finding a venue which will meet all the criteria and is located in a reasonably convenient position for the considerable numbers of members living in south Essex while not being too long a journey for other members who are scattered across the rest of the southern part of our region. We know members in the northern part are well served by the Norwich meeting but if any of you feel you can make it to this part of the region as well, we would love to see you.

Hilary Croydon

Birmingham Area

Hi Everyone,

We are thrilled to be able to tell you about a Midlands HSP Support group, that we have started.

A few of us met at the AGM in June 2014, and after exchanging emails, 7 of us met up in early October in the south of Birmingham, with a view to starting up a Midlands HSP Support Group.

We were hoping to have a second meeting booked at a new venue in plenty of time to publicise in this Newsletter but although we have venues in mind we haven't quite had time to finalise plans.

We are hoping to hold this second meeting sometime in April and we'll communicate by email to all in the region.

Please feel free to contact me:

pennycohen57@hotmail.com to either let me know you'd be interested in attending or to find out more.

Penny Cohen

Region 9 - St Helens – Sat 25th October 2014

The Region 9 meeting was very enjoyable. We were chatting and socialising so much, that when Dr Siva and Alison were ready to start, it took some doing to get people to calm down a bit. The speakers stayed till 3:30 ish, so we had plenty of time to continue afterwards. Some of us stayed for a meal at the end and enjoyed that.

We were joined by Dr Siva Nair, Consultant Neurologist and Alison Clarke, Clinical Physiotherapist from Sheffield.

Dr Siva talked about HSP and answered various questions. There are now 44 different “types” of HSP, differing in exactly which part/parts of the nervous system is affected and what type of inheritance it is passed down through.

The research into HSP is mainly centred on genetic research, i.e. the causes of HSP and little on treatment or cures. In the last 10 years there have been 300+ research projects in HSP genetics and only 11 on treatment. In comparison there were 200 times more research projects on treatment of Multiple Sclerosis. Dr Siva suggested that we, as a group, perhaps need to be little more vociferous in influencing research projects. It is not an easy process, he acknowledged.

However, the funding for research is changing and priorities are also about to change. The Government are encouraging “user control of research”.

The use of nasal cells for repairing damaged nerves was also raised, following news of such treatment for spinal injuries cases. Dr Siva said it was early days yet and at present he did not feel confident that it would work on HSP, although some research into this topic in relation to HSP is being carried out in Australia.

In relation to the prevalence of HSP, Dr Siva said that countries closest to the equator have the lowest incidence of HSP and as one moves further north the

incidence increases. Generally there are between 4.3 – 7 cases per 100,000 of population.

A question was asked about the possible benefits of SATIVEX. SATIVEX is a cannabis based spray, licensed in the UK for use by Multiple Sclerosis patients. Its main effect is on relieving spasms and spasticity. Apparently it is not licensed for the use of HSP patients. The process of licensing has to involve trials and since HSP is such a varied diagnosis, this has not been done. Enquiries have been made to the MHRA (Medicines and Healthcare products Regulatory Agency) as to why HSP patients cannot use it, if it deals with the same symptoms (spasticity and spasms). Apparently about 1/3 of MS patients benefit.

There was some discussion of the use of Baclofen and Baclofen pumps. In Dr. Siva's opinion this drug was more appropriate for patients who are wheelchair bound. For HSP sufferers who are still walking, this may pose problems with getting the dosage exactly right and side effects. There have been 3 studies on the use/benefits of Baclofen pump. Only 2% of users in these studies had no side effects and a big question remains as to whether it helps with walking.

Alison Clarke stressed how important it is for HSP sufferers to maintain a range of movements (arms, legs and the trunk) and not concentrate solely on the lower limbs. Muscles need to be co-ordinated for movements to work well.

The Sheffield NHS have a specialist Spasticity Management Clinic, there are others e.g. in Birmingham. The clinic deals with general exercise advice, can prescribe Botulin, Baclofen and Baclofen Pump, FES (Functional Electrical Stimulation). Botulin (saline and Botox) injections can be used for a targeted treatment. The dose and follow up treatment are really important.

Alison also suggested that hydrotherapy was very good for HSP. Many pools have special sessions where the water is heated to

a higher temperature and exercises can be carried out more easily in the water as the body does not have to support all the weight.

She acknowledged that access to physiotherapy may be limited and sporadic in different parts of the UK. However, if we experience any changes in movement, we should ask for a referral to a neuro specialist physiotherapist who has an understanding of HSP, straight away.

The Chartered Society of Physiotherapy is a good source of information about NHS and private Physiotherapist specialisms.

And we have all probably been told this many times before, **BUT when walking the heel should touch the ground first.**

Irena Pritchard

Forthcoming Events

Region 5 Meeting

Saturday March 14th 2-6pm
The Oaks Restaurant
Premier Inn, Norwich Airport
Call Barbara Jones on: 01603 423 267

Region 9 Meeting

Saturday March 14th 2-6pm
Meeting Room
St Helens South Premier Inn
Eurolink, Lea Green, St Helens, WA9 4TT
Call Irena Pritchard on 01524 261 076

Region 4 get together

Saturday April 11th 2pm onwards
The Dartmoor Lodge Hotel, Ashburton
Call Ian Bennett on: 01202 849 391

Afternoon tea Regions 1 & 2

Saturday April 18th 3pm – 6pm
The Clockhouse Milford
Call Jane Bennett on: 020 8853 4089

HSP Support Group AGM

Saturday June 20th
Trident Technology and Business Centre
Leamington Spa

Region 3 Meeting

August 15th 2015 1.30pm – 4.00pm
The Orange Tree Public House
100 Stevenage Rd, Hitchin, SG4 9DR
Contact Della Brookman: 07710 637 941

New Members

We welcome the following new members

David Cardwell
West Yorkshire
Region 10

Dorothy Townsend
Blackburn
Region 9

Ian O'Neill
Glossop
Region 9

Jenni Preston
Hornchurch
Region 1

Louise Sumsion
Bristol
Region 4

Neil Jackson
Mansfield
Region 8

Paul Eccleshall
Birmingham
Region 7

Sally Meikle-Janney
Huddersfield
Region 10

If you are interested in contacting the new members listed, please contact the relevant area co-ordinator, or the membership secretary, who can then supply the necessary contact details
