



HSP Newslink

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

Issue 33 – September 2014

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

Fundraising the Old Thatch way



Left to right Standing, Gemma, Carol (pub landlady), Tanya and seated, Carol Parsons, Sarah and Pete

Jason bought his girlfriend Gemma a skydiving experience and the HSP Support Group are now well over £1500 richer as a consequence. Let me explain:

I got to know the Roberts family some time ago when our daughters all attended the same school. Sarah and Pete Roberts are mum and dad; Gemma and Tanya are their daughters, both of whom work behind the bar at the local pub which is called The Old Thatch, located between Ferndown and Wimborne in Dorset.

When Gemma discovered she had a skydive to look forward to, the other girls in the family, Sarah and Tanya thought they'd like a piece of the action so they also signed up for the jump. During discussions the girls decided that they wanted to raise money for a good cause and Sarah contacted me to see if I knew of any worthwhile, deserving charities. The rest is history! They particularly like the fact that **every penny raised will be put to good use** and not absorbed by excessive administration costs and wages.

The staff at The Old Thatch have been brilliant and offered their full support. They have HSP collection boxes behind the bar and they recently held a BBQ with live music to raise further funds. Carol Parsons, another of the bar staff from the pub decided that she'd also like to jump and she has been busy gaining sponsorship. All the girls who are jumping have pages on JustGiving.

Further discussions took place between the Roberts family and the pub management and they agreed that they'd like to support a Dorset resident affected with HSP. They're hoping to purchase mobility equipment that could have a big positive impact on a local individual's quality of life. Consequently I've written to all Dorset members explaining the fundraising and I've invited them to make me aware of any equipment that may be useful.

Tanya used to work at the Garden Centre that is located a couple of hundred yards down the road from the pub and when I explained to them how she's fundraising for HSP they agreed to have our collection pots at their checkouts. They particularly liked the fact that it was for a local cause.

Gemma has organised a charity car wash in the pub car park, charging £5 per vehicle with all takings going in the pot.

The parachute jump was originally booked for August 16th and there was great excitement all around. Unfortunately the weather wasn't suitable for the jump to take place so it'll be re-arranged to be performed as soon as possible. The delay could be a blessing in disguise providing the opportunity to raise even more.

It's been a pleasure to be involved with this fundraising. Every time I've popped into the Old Thatch to empty the collection pots, I've stayed in the bar for an hour or two getting to know the locals and consuming plenty of fine ale. The things I do for HSP! We're hoping to organise a special evening in the pub in the not too distant future when the lucky Dorset HSP

member will be presented with his/her new equipment.

All the people involved seem to particularly like the fact that they are helping a local individual. Maybe other HSP members around the country can get other organisations interested in similar fundraising, targeting someone in the county who needs some help.

Huge thanks to everyone listed below who's been involved with this

Sarah, Pete, Gemma and Tanya Roberts

Carole the landlady of the Old Thatch

Carol Parsons who works behind the bar

All the staff and regulars at the Old Thatch for their generosity and support

Wimborne Garden Centre

Jason who had no idea what he'd initiated when treating his girlfriend Gemma to a skydiving experience.

It's been fun!!!

Ian Bennett

Chairman's Column

Although only three months have passed since our AGM it seems longer than that to me. The minutes of the AGM and a summary of the presentations are all included in this edition. I can't thank Adam Lawrence enough for his hard and thorough work in summarising the presentations so well. This is invaluable for members who were unable to attend and provides a good reminder for those who were present. Adam's input has made this issue the longest issue ever but it's far too interesting to not be included.

I've been delighted to hear how many members have benefited from what Sportability has to offer as a result of learning about them at the AGM. Sportability activities that our members have recently participated in include quad biking, archery, gliding, shooting and even water skiing.

Summer has been and gone since the AGM but what a great summer it was. Loads of sunshine and I've spent many hours outdoors enjoying it. I've recently experienced my annual visit to the Bournemouth Air Festival. This year I was interviewed and the footage went out live to the big screens on the beach. I was raising awareness of the Flying Scholarships charity but I managed to discuss HSP during the interview. In July I went to the Royal International Air Tattoo for the Flying Scholarships presentation ceremony, the highlight of which was witnessing Leanne Piccirillo being awarded the Wings Around the World trophy. This trophy is awarded annually to the female who was seen to have got the most out of the flying scholarship. Many congratulations Leanne.

My new scooter which I've discussed a little in the previous two issues has had loads of use getting me out and about in the sunshine this summer. I now don't know what I'd do without it.

On the subject of mobility scooters, it's worth highlighting one particular model. You'll notice that two of the members' letters included in this newsletter discuss the Monarch Mobie lightweight scooter. Both individuals speak extremely highly of it so if any of you are considering a new lightweight scooter, this model has to be worth looking into. Thank you to John and Hugh for sharing this with us.

The fundraising at my local pub which you'll have read about on the front cover is just one of many recent money raising schemes for the HSP charity. Michael Kelleher has done an amazing job cycling 50 miles for us and raising over a thousand pounds in the process. Michael has written a few words about this (page 4) and on behalf of everyone involved with the group, I pass on our thanks for such a great effort. I met Michael a couple of months before the event and taking his mobility into consideration, it was certainly no easy task. When you read his words, you'll realise just how much he put into it.

The ice bucket challenge which has been a lot of fun has raised significant funds for us and I've certainly enjoyed watching many of you getting a good soaking on Facebook. My turn had to come and wow it was cold! Thank you to everyone who has been a part of raising some money for us recently, however you've achieved it.

On the subject of finances, the committee are shortly getting together to decide which researchers we'll be supporting this year. The outcome of this will be detailed in the next edition.

I think I've mentioned in previous issues and I certainly brought it up at the AGM that we now have a database listing members who are happy to volunteer their time to help research. This doesn't mean that you are committed to helping but it gives the researchers a list of people to approach. If this is of interest, please let me know. Helping researchers is one of our most important activities.

It is very pleasing to see that we have increased the number of meetings we hold around the country enabling more members to get to know others affected with HSP. You'll notice the list of meetings in the 'Forthcoming Events' section is getting longer. Thank you to Hilary Croydon, Barbara Jones and Irena Pritchard for all your help and enthusiastic support with this very important activity. Our Facebook group; Hereditary Spastic Paraplegia's Unite' has recently achieved a milestone. It has now exceeded 500 members which is considerably more than the Support Group. If you're not a member of this facility, please join up as there is always plenty of activity going on.

Please keep the interesting Newsletter material coming in. This edition is the longest ever, partly because of the articles and letters I've received from our readers and members.

As always I look forward to seeing many of you at future meetings, the first of which for me will be Ashburton on October 18th.

Ian Bennett

Michael's Fundraising

Cycling 50 Miles to Support Spastic Paraplegia for Hereditary Spastic Paraplegia Support Group

This event took place as planned Sunday 22nd June, perhaps one of the hottest days of the year.

However 9am we set off from Deal pier in the heat with a gruelling 50 miles ahead of us. The first stage was to follow the coast; we did, through and over one of the finest golf courses, across several fields and fences/gates etc, until it was obvious the leader didn't really have a clue. So lost at the start, over an hour wasted we turn around almost back to the start. Thank God for modern phones with sat nav's. Back on route we proceeded around the coast through several towns and villages.



Michael on route

Sandwich, Ramsgate, and then Margate our first stop to top up with fluids and refreshments. What a backup team, now well behind time and several miles done unnecessarily we proceeded along coastal roads, through field treks towards Reculver towers (stop 2). It's such a wonderful relief when greeted with more refreshments and 5/10 minute break as the stragglers catch up. Several more stops were taken before finally reaching our destination, changed at the last moment on route due to the pub kitchen burning down that same morning. So a new pub pre-warned by our backup team.

They looked after us all with a secure place for our bikes and refreshments in (beer) and food. However just to top things off, at 5pm I collapsed in the beer garden. The colour disappeared from my face, I started to shake, then gone!!!!. Within seconds the first response medical team arrived followed closely with the ambulance and paramedics. Fully checked over within the hour I recovered. They put it down to total exhaustion. But we did it, Fantastic! Raising more than my set target of £500. The total now paid in is £1,022.

Michael Kelleher

Situations Vacant

The committee believes that it is important to our members to meet others with HSP.

We have the AGM of course but not everyone has the stamina to make long journeys.

So we are keen to encourage meetings around the country.

Some meetings have been running for 10 years (Milford and Ashburton) and have regular attendees who have become good friends.

Last year Ian and John set up new meetings in St Helens and Norwich. But there is more to do and we would like to have a new person doing it.

What they have to do?

Firstly set up new meetings. That can be done by looking at the map of where our members live (yes, we have one!) and seeing where there are gaps in our coverage.

Secondly select a venue and a date. We use Premier Inn quite a lot as there is accommodation and a restaurant, both usually accessible, and sometimes a meeting room. The committee (i.e. John) can help by contacting everyone within an hour's drive and publicising the meeting. We can also help by paying for the venue hire.

Thirdly encourage someone local to take over the meeting as they have the important local knowledge.

Finally, and this is for the longer term, encourage the local organisers, collect feedback and distribute good ideas, and encourage raffles and the like to make the meeting self-funding.

It needs someone who is happy to talk to people and encourage them. Could you do that? Ian would be pleased to hear from you.

The meetings above are generally attended by us oldies and may not be much fun for younger (15-25). If you are in the younger age group would similar meetings be of interest to you if they were restricted to this age group?. It may be that there are more modern methods of communication that you prefer eg Facebook, Twitter etc. So it would be useful to get your views Please let us know what you would like and we will see what we can do.

The HSP Committee

Research Column

Many people have seen or heard of the ALS Ice Bucket Challenge. I watched this through Facebook and it circled closer and closer to me with friends and family being challenged and then nominating on.

I had spotted that ALS was a mis-diagnosis for HSP in my survey last year, and so I thought about the similarities and differences between them.

So, I start with a definition:

Motor Neuron Diseases (MND) are a group of neurological disorders that affect motor neurons in adults and in children. Motor neurons are specific types of cells that control voluntary muscle activities such as speaking, walking, and breathing.

http://www.alsconsortium.org/motor_neuron_disease.php

Motor Neuron Disease can refer either to the most commonly occurring form - ALS (Amyotrophic Lateral Sclerosis) also known as Lou Gherig's disease, or to the broader spectrum of conditions.

Everyone who has looked up HSP on the internet must have read "Hereditary Spastic Paraplegia (HSP) is a group of rare, inherited neurological disorders". HSP is a neurological disease because the long nerves in the spine are gradually degenerating - the nerves are the neurological system (paraphrased from [HSP Research Foundation](#)).

HSP

The one paragraph description of HSP (with parts borrowed from both previous links) is:

The primary features of HSP are spasticity and weakness in the legs, varying between individuals. There is progressive difficulty walking and symptoms worsen over time. Initial symptoms are typically difficulty with balance, stubbing the toe or stumbling. Changes begin gradually. As the disease progresses, canes, walkers and eventually wheelchairs may become needed, although some people never require assistive devices. A wide variety of symptoms are observed across cases and over time, including balance, fatigue, bladder and back pain. The majority of individuals with HSP have a normal life expectancy.

ALS

Whereas ALS describes as (NHS: <http://www.nhs.uk/conditions/Motor-neurone-disease/Pages/Introduction.aspx>)

The symptoms of motor neurone disease begin gradually over weeks and months. Common early symptoms are: weakened grip, weakness at the shoulder, "foot drop", dragging of the leg, slurred speech. As damage progresses, symptoms spread to other parts of the body and the condition becomes more debilitating. Eventually, a person with motor neurone disease may be unable to move. They may also find communicating, swallowing and breathing

difficult. The condition is not usually painful. ALS is a severely life-shortening condition for most people. Life expectancy for about half of those with the condition is three to four years from the start of symptoms.

PLS

Another form of motor neuron disease is PLS - Primary Lateral Sclerosis.

http://www.ninds.nih.gov/disorders/primary_lateral_sclerosis/primary_lateral_sclerosis.htm

Primary lateral sclerosis (PLS) slowly gives rise to progressive weakness in voluntary muscle movement. The first symptoms are often tripping or difficulty lifting the legs. The disorder often affects the legs first, followed by the body, trunk, arms and hands, and, finally the bulbar muscles (muscles that control speech, swallowing, and chewing). PLS progresses gradually over a number of years, or even decades. Life expectancy is normal.

Summary

I won't go further and list any other types of motor neuron disease, but note the similarities:

- Early symptoms for all three are similar.
- All three cause upper motor neurons to degenerate (those from the brain down into the spine)

And the differences:

- ALS and PLS progress to more parts of the body
- Life expectancy for ALS is reduced.

It depends where you read to see if HSP is included within the motor neurone disease list or not. I certainly found some websites which did and others which didn't.

I take the view that all this ALS ice bucket challenge activity raises awareness of diseases caused by degeneration of the nervous system, and that with all such

causes it is down to the participants to personalise their message to encourage others to donate money. On this basis when I was nominated I donated to the UK HSP Support Group, and several friends and family who were nominated at around the same time did so as well.

If there are any readers who were not lucky enough to be nominated for the ice bucket challenge and would like to make a donation, then I encourage you to do so, even if you don't empty a bucket of icy water over your own head. If you're stuck to find a relevant charity which you would like to donate to, then why not choose the UK HSP support group?

<http://www.hspgroup.org/>

You can read Wikipedia's ice bucket challenge story here too:

http://en.wikipedia.org/wiki/Ice_Bucket_Challenge it would seem that golfer Chris Kennedy was the first to nominate/donate for ALS on July 15 2014, and former basketball player Pete Frates who has ALS and is a patient advocate began spreading the ALS word through Twitter.

Survey on medication & exercise

Last year members may recall I surveyed about mobility, symptoms and mis-diagnoses. I am undertaking a survey this year, focussing on medication and exercise which I would welcome members to spend a few minutes completing. People are welcome to provide as little or as much information as they want. I will collect answers for the rest of the year and report on my blog 28th Feb 2015. The link to the survey is:

<http://www.surveygizmo.com/s3/1797674/HSP-Medication-diet-and-exercise>

Adam Lawrence

Useful Information

Toilet Talk

Constipation and bloatedness are two unpleasant factors that many people with HSP contend with on a daily basis. Here's an interesting product that could help this problem.

The 'Squatty Potty' is a simple but very cleverly designed piece of equipment that could help with some of the HSP related bowel problems.



As can be seen it's simply a platform that enables the user to adopt a squatting position which is a more natural way to 'do your business' and tends to involve less straining. It's a great way of overcoming constipation and can generally make the whole process much easier.

The 'squatty potty' is not expensive and I was delighted to discover that it's available in the UK. I contacted a supplier and they have very kindly offered a 10% discount to our members. To order or for further information, visit:

<http://www.stressnomore.co.uk/brands/squattypotty.html>

To qualify for the 10% discount simply quote **HSPgroup10** within the gift code field while on the 'shopping basket' page when placing your order.

HSP Group Funds

Funds are now 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.

Members' Letters

Dear Ian,

Following my letter printed in the last HSP Newsletter I said I would update how I got on travelling with my mobility scooter. Just to recap I wanted a small scooter that was easy to take away, fit easily in the car and be available for shopping as opposed to a full size model.

The choice for me was between the Luggie or the Monarch Mobie, both of which compact to about 17in x 17in x 20in and weigh around 50Lbs (23Kgs), and I purchased the latter.

At Easter Stella and I went to Cyprus with Thomson for 7 days to celebrate my 60th birthday, having booked the holiday just a few weeks before. I was able to ride the scooter right up to the cabin door going out from Stansted and on the return flight and also it was available to me on leaving the cabin at both ends with assistance at all points. The only problem was that they would not take the scooter on the coach due to safety with regard to the lithium battery so I had prepaid for a taxi from the airport to Hotel and return journey.

Getting around the Hotel and grounds presented no problems at all and it was easy to use outside in the street.

This was a trial run as in January we had booked a two centre holiday in the Caribbean, and as it proved a success in Cyprus, we arranged to include the scooter as part of our plans. At Gatwick they took

the scooter just before the jet bridge and arranged wheel chair assistance on to the plane. In Barbados it was waiting for me on the tarmac and we were assisted to the check-in desk for a connecting flight, then to the 19 seater plane to Bequia (Bekway). On leaving Bequia I was able to ride up to an 8 seater plane to Grenada. The pilots on both these small planes were fascinated by the scooter and couldn't believe how small it folded down. At both Hotels there were no problems in getting around, although we knew beforehand that it was not practical to take it outside the Hotel in Bequia. On leaving Grenada for home I rode up to the plane steps and was then carried up in a chair by 3 airport assistants. The only hiccup came at Gatwick as flight or ground staff didn't know where my scooter was so we had to wait on the plane while enquiries were made. Eventually it was found near the baggage reclaim point after I had got there by wheelchair and buggy assistance. In all places people were amazed by the scooter and in Cyprus particularly we could have sold it many times over. It has been a good buy.



Monarch Mobie folded for the boot

P.S. I sent an email to Monarch letting them know how well I had got on with the scooter entitled 'Mobie on vacation' and they contacted asking if they could use my photo for advertising. So to my surprise a couple we got to know over a few days in Grenada during June sent an email to say they had spotted my picture, supplied by Monarch, in the Mail on Sunday 10 August

as part of a small article headed 'Scoot further than you'd think'.

John Patching, Tiptree, Essex

Dear Ian

We always enjoy reading Newslink which we find both interesting and informative. I was interested to read in the last issue John Patching's letter in which he states he has bought a Monarch Mobie folding scooter.

I bought one last summer and we have found it to be invaluable. We took it with us in the car on a holiday to Wales; in September I took it with me to travel by plane to St. Petersburg, where it was invaluable for going round the Hermitage museum and other royal palaces. ('tho it needs to be said Russia is not disabled access friendly) More recently, we went to visit family in Italy and took it with us by plane and were able to use it going along the seafront and visiting the Capodimonte Art Gallery in Naples. We have not yet taken it with us on a train, but hope to do so soon. It is easy to fit into the boot of the car when we go into the town centre on a shopping trip.

The great thing about having a Mobie at airports is that you can stay on it right through to boarding, and get on it again soon after arriving, and still have assistance. I can't recommend one too highly. The main problem is they are expensive, about £1500 -£1600 – twice the price of an average scooter, but worth every penny.

Yours sincerely,

Hugh and Mary Beavan

Dear Ian

We really enjoy reading the many exciting stories in the HSP Newslink. It is good to hear of the fighting spirit that many people with HSP seem to have. We have enjoyed flying planes, hot air balloon rides, etc. in the past. Now we are no longer spring chickens, but we are still managing to find

new things that we haven't done before and this is our story of our **'First Auction'**

We sorted out a little pile of things that we no longer required and listed out the items. Instead of the local charity shop or car boot sale, we decided to use an auctioneer and valuer. We looked up on the internet for auction dates and noted that valuations are done 9-11.00 a.m. each morning Monday to Friday, so off we went with our collection and our list. Of course, Murphy's Law stepped in and the valuer was away on holiday when we chose to go. We weren't expecting any great fortune, so we decided to leave our things for the sale in two weeks time. The lady told us that the auctioneer would look through them and assess if anything should be put separately in the sale, she told us he had a good eye for picking out special lots. The viewing was one day before the sale, but as we were selling and not buying, we decided just to go on sale day. On arrival we were handed a list of our goods for sale taken from the catalogue and each had a lot number. Some items had been listed singularly. The receptionist told us that they sold approximately 120 lots per hour, which meant we were able to assess when our lots would be up for sale. Being in a wheelchair, we were shown to an area where the wheelchair could fit and the auction began. At 120 lots per hour, I thought we would soon lose touch with what was happening, but it was speedy, but clear and before we knew it, our first lots came up for sale as trays of items.

First - £5

Second - £12

Third - Some old corkscrews (a joke gift of 50 corkscrews from friends on Terry's 50th Birthday) made £48

Fourth - £1 - Maybe the car boot would have been a better option!

It looked as if we may get around £100 - Terry's guesstimate before we came. Still we were enjoying the atmosphere.

After these four lots, we calculated that there was a break before our next lots came up for sale, so it was time to enjoy the café. It was a glorious day, so we took our food and drink out into the sun and lots of chat with people who were buying and selling meant time flew and we were back inside before our jewellery bits and bobs (broken watches from Mums and Dads, etc – nothing special). The auctioneer had spotted something about two of the items and these were listed as individual items.

£65 for an old broken fob watch of Mum's – wow!

The next lot went for £95, another for £100 and then for £140!

Then four boxes of old cigarette cards made just £55, but some old black and white Beatles cards which were of poor quality and had been acquired free with Sun newspaper and chewing gum packs had been listed separately from the cigarette cards, these started at £10 which rapidly went to an unbelievable £80

All in all we made over £600 and had a really good day out. Terry was pleased that I didn't have a paddle to bid, otherwise we may not have gone home with any money. What do I mean – may not! It would have been a certainty.

Perhaps we will try buying next time! I can see Terry turning pale at the suggestion.

We would like to thank Byrnes Auctioneers for their helpfulness and friendliness of the staff.

Our final comment is that we would recommend you give it a go. A great fun day.

Pat and Terry Reed Region 9

Dear all,

In overcoming our physical battle I have sensed first meeting the ongoing challenge of becoming depressed by it by finding encouragement from progressively extending our capability as independently as safely possible, as we so very gradually extract more from the functioning parts of

our imperfect bodies. I have been very pleasantly surprised by what can be achieved by the upper body when compensating for the virtually dormant Shank's Pony and coaxed, with the help of elbow crutches, to confidently carry me up and down stairs and slowly over uneven terrain. Surprising those who think I should not even dare to try such exploits is, I've found rewarding, insofar as it broadens their awareness of what we remain capable. Our carefully controlled efforts inspire them more readily to be helpful, if possible, and our exclusion from their activities is hopefully not so readily assumed.

I took to elbow crutches five years ago if only to avoid damaging the cartilage in my stick-wealding shoulder, initially allowing my body weight to continue resting on my feet as I shuffled along. Then, with the prospect of some very appealing foreign travel almost a year ahead, I found that by lifting myself bodily and swinging it through between my crutches much improved my 'walking' pace. I was professionally assured that provided the initial upward lift was achieved painlessly, this was safe to continue. All the shoulder muscles newly coerced into this demanding action necessarily went 'into training' affording me as the weeks and months passed, the capacity to move further between each breather. In parallel with this came a strength, if not dexterity, to reliably 'drop myself' where and when I wanted without imbalance. Tumbles may happen largely as a result of not seeing, or reacting fast enough, to an obstruction to the movement of me and/or my crutches – maybe simply going too fast! To have 'where there's a will there's a way' as a motto is highly desirable, but recognition of having exceeded the bounds of this dexterity must of course have its place in the interests of safety. Gradually improving one's upper body strength is therefore very rewarding. In Issue 30 I wrote of my exploits in arctic Norway early last year in a failed attempt to see the Northern Lights. Continuing to thrive then on unusual experience, 'where

next?' was the inevitable question – somewhere within our British Isles which a good many have never heard of and it completes the sequence 'Muckle Flugga. Lowestoft. Alderney'. Being the northern, eastern and southern extremities of our islands. Ignoring Rockall, the St.Kilda archipelago (inhabited only by the MOD now) is the western one; a world apart that is a haven for roughly a million sea birds, which until 1990 provided sustenance for the longstanding community on the largest island, Hirta. So I started looking into the prospect of going to possibly the most isolated part of our country and to 'pole' down the one-time village street, the remains of which are now in the care of the National Trust for Scotland. A boat trip each way of over three hours across 41 miles of Atlantic Ocean west from the Outer Hebrides, sandwiching a stay on Hirta of roughly four hours, lay in prospect, all critically subject throughout to the unfettered elements. Were these aspirations themselves unfettered, let alone the weather and getting ashore via a Zodiac inflatable?? I clearly had no idea.

I said, in closing last time that the strength left in our upper bodies unaffected by the paraplegia can gradually be made to achieve so much more, tethered only, I hope, by our Maker. He knew only too well I wanted to go, but, as with Norway, I left it with Him, knowing that I should safely go solo only if all the circumstances were in His judgement right. From all I'd read, I would not have been too disappointed, perhaps even relieved, if all my outline planning came to nothing. A friend with access to good meteorological data posted a favourable to me and I somehow felt it right to set the whole circular tour, via Ullapool, Skye and Arran, in motion at less than two weeks notice. Even then, I was treated as being on 'standby' for a sailing on Tuesday and, failing that, on the following Thursday – I might still never make it. I learned further, after the event, that reaching Village Bay, Hirta did not automatically ensure being able to go

ashore. On the Monday evening, having met folk who had just returned from a misty encounter with the island, I learned somewhat unnervingly that I was indeed going the following morning at 8am.

I may have been seasick three times on the outward trip and had modest difficulties getting ashore, but that potentially volatile weather was consistently ideal so that my venture on the island, keeping carefully off the boggy ground, gave me such an immeasurable experience. Our Maker knew precisely how to temper my enthusiasm so as to grant me that something, forever between my ears enriching my state of mind, that testifies, as before, to his ongoing care.

John Moule Region 8

Regional News

HSP Region three meeting.

Our regional meeting was held, as usual at The Orange Tree Pub, in Hitchin on Saturday 16th August.

The meeting was well attended. In all, there were twenty people who came along for a relaxed, informal time, to get to know each other, have a few laughs and chat about experiences living with HSP and other topics.

Ian Kitchen arranged for his Consultant Neurologist, Dr Wilkinson, to come along to answer any questions on HSP that members might have. Dr Wilkinson has worked with HSP in his research, for qualifications and now sees Ian Kitchen and other neurological patients and specialises in Botox injections. A lot of questions were fired at him and he answered each with understanding. Explaining that there are over 50 known genes and other interesting information. We thank him so much for spending an afternoon with us. Which was appreciated by all.

I asked for feedback from members, to let us know how they thought the meeting went and how, if needed, it could be

improved. I received this e-mail from a new member, who telephoned the previous evening before the meet and came along to join us.

Hi Della

I've lots of positive thoughts for you.

I received a lovely welcome (and thank you both especially for that) and was included straight away in conversation with yourselves and other members. I learnt a little about the Support Group as a whole, about the individual branches and of the use of Facebook to share information, support and conversation. I was impressed that members could apply for funding to help their mobility in and outside the home. A glass of wine and some lunch made me feel even more included. Having a Neurologist join us to share thoughts and information was an added bonus.

Before joining you on Saturday Della I didn't know that there were different types of HSP and it did make me sad to see how it had affected the mobility of the two youngsters especially.

That's all my input about the meeting. I hope some of it is of use to you.

Keep happy and positive and see you again later in the year.

All the best' Alayne

This is a very positive response and Ian and I thank Alayne for coming along. I am glad I did not frighten her off, after our conversation the evening before! :D.

Also, one of our regular attendees, Hilary Croydon, has arranged a meeting on 21st September 2014 at Holiday Inn, Abbots Lane, Eight Ash Green. Colchester. CO6 3QL. 1300-1500. Hoping this will become a regular HSP meeting.

Roger and I are planning to go along and hope others in the area can too.

Small meetings around the UK are becoming more popular and are a great way to keep in touch and get to know fellow HSP'rs to keep up with news etc.

We are trying to arrange a Christmas lunch, possibly in January and will keep you posted as and when we have put something together.

Della Brookman & Ian Kitchen.

Region 3 Coordinators.

Forthcoming Events:

Region 3 Meeting

August 15th 2015 1.30pm – 4.30pm
The Orange Tree Public House
100 Stevenage Rd, Hitchin, SG4 9DR
Call Della Brookman: 07710 637 941 or
Ian Kitchen: 07540 476 735

Region 4 get together

Saturday October 18th 2pm onwards
The Dartmoor Lodge Hotel, Ashburton
Call Ian Bennett on: 01202 849 391

Colchester Meeting

Sunday September 21st 1-3pm
Holiday Inn, Colchester, CO6 3QL
Call Hilary Croydon on: 01284 728 242

Region 5 Meeting

Saturday October 18th 2-6pm
The Restaurant
Premier Inn Norwich Airport
Call Barbara Jones on 01603 423267

Afternoon tea Regions 1 & 2

Sunday November 23rd 3pm – 6pm
The Clockhouse Milford
Call Jane Bennett on: 020 8853 4089

Region 9 Meeting

Saturday October 25th 2014 2–5pm
Meeting Room
St Helens South Premier Inn
Eurolink, Lea Green, St Helens, WA9 4TT
Call Irena Pritchard on: 01524 261076

HSP ANNUAL GENERAL MEETING Saturday 21st June 2014

Minutes

Apologies for absence

Apologies were received from Tony & Angela Barnicoat, John & Sonya Mason, Simon Hubbard, Mixhael and Ann Horsman, Jane and Michael Bennett, Maurice Charge, Hugh and Mary Beavan, Mark Tomlinson, Emma Saupé, Sherrill Casburn, John and Carol Moore.

The Minutes of the 2013 AGM were agreed to be a true record: proposed for acceptance by Debbie Best and Ronnie Ferguson.

Committee reports:

Stephanie Flower welcomed new members to the meeting and hoped to be able to talk with them before the day's end.

Ian Bennett reported on meetings being "fostered" in the North West and in East Anglia. He was now hopeful that these could be continued by local members. Another meeting had been arranged by Lorraine Saupé on the South Coast, in Bournemouth. She had hired a "disabled friendly" beach hut for the day and a dozen members had met there. Ian was hopeful this would also continue as a regular event.

Ian thanked members for their interesting contributions to the Newsletter and requested that members continue to submit content to help ensure future editions are an interesting read.

David Harris presented the Treasurer's Report on behalf of John Mason, Copies had been handed around to all attendees. John's notes and observations were read out including thanks to Sonya Mason for her assistance through the year and to Theo White as Auditor. After an opportunity for questions, adoption was proposed by Ronnie Ferguson and seconded by Andre Wheeler. This was approved by the meeting with a show of hands.

Election of Officers:

Chairman, Ian Bennett was proposed by Maggie Gillson and Seconded by Gill Ransome.

Treasurer, John Mason was proposed by Terry Duffy and seconded by Della Brookman.

Secretary, Dave Harris was proposed by Barbara Kevan and seconded by Phil Burton.

Committee Member Simon Hubbard was proposed by Ronnie Ferguson and seconded by Debbie Best.

Helpline, Stephanie Flower was proposed by Gill Ransome and seconded by Barbara Kevan.

Committee Member John Moore was proposed by John Flower and seconded by Caroline Begg.

All the above appointments were approved by the meeting.

Any Other Business. No items were raised.

A Summary of the AGM Presentations

HSP Research at Plymouth University - Prof Jon Marsden

This article covers both of the times which Jon Marsden spoke at the AGM. He spoke first to introduce Plymouth Universities areas of research interest, and to introduce his colleagues. Later in the day he suggested some research which he would like to commission..

Plymouth University Research

Jon began by defining HSP into two parts, being the two main areas of research. The "H" - hereditary - is the genetics side of things, and research seeks to understand what causes HSP, what the different affected genes do, and that kind of thing, and seeking potential treatments.

The "SP" - spastic paraplegia - is about the effects, and this is what is concentrated on at Plymouth. This research seeks to understand what symptoms are seen, how these symptoms affect quality of life, and how the symptoms can be reduced.

Jon observed that there is a third element, which combines the two parts looking at overall service delivery and support, which would consider things like physiotherapy, mobility aids, and so on.

At Plymouth, over the years, the team has been looking at the impact of HSP, understanding what the effects are. They have undertaken research by asking people questions and undertaking examinations and measurements. Measurements might include stiffness or strength, or sensitivity to vibration, and these would be looked at in the context of the study being undertaken - for example, how does HSP affect people balance.

In this regard, HSP affects hip muscles, which limits sideways leg movements, and stiffness in ankle muscles affects back and forth movements, so the HSP gait involves swaying from side to side and a lot of dragging feet along the floor. But, some stiffness is needed for balance, and the contrast is that if you reduce the stiffness to increase mobility then you may decrease balance. A certain amount of stiffness is needed in our muscles to allow balance normally, and HSP adds to this stiffness. This element can be treated with stretching, and some of the work at Plymouth has been to target therapies to the symptoms.

Looking at the overall service delivery side of things, Jon reported a study that they have recently completed which examines this process, and concludes that "People with HSP require better self-management advice, information and support."

<http://www.plymouth.ac.uk/pages/view.asp?page=32853>

<http://www.ncbi.nlm.nih.gov/pubmed/23944176>

<http://informahealthcare.com/doi/abs/10.3109/09638288.2013.824032>

In Jon's presentation later in the day described this research. One of their findings is that there is a poor evidence base out there on which to make decisions - either about providing treatments to people or about funding further research. Jon listed the common treatments which have been identified as providing a benefit by people with HSP:

- Genetic counselling
- Anti-spasticity drugs
- Physiotherapy
- Occupational Therapy
- FES (Functional Electrical Stimulation)
- Tai chi / Pilates / Acupuncture
- Hydrotherapy

There are limited studies covering some of these, perhaps 50 papers in total covering all of these.

Jon reported that the key issues for people with HSP in the south west of the UK (Devon and Cornwall) were:

- Length of diagnosis
- Lack of publicity/knowledge
- The need to travel to get access to specialist knowledge (e.g. travel to London)
- Allocation of services (i.e. availability of funding for investigations/treatments)
- Co-ordination of care (healthcare professionals talking with each other)
- Lack of evidence for treatments/interventions

"Telehealthcare"

Jon then described a potential model for the provision of specialist HSP care, drawing on existing practice in other areas. (I'm not 100% sure it was called Telehealthcare, apologies if not) Essentially, satellite centres would be set up to provide specialist HSP care two times per year. Within these centres there would be video-conferencing to specialist consultants, and there would be a range of other support available during the day.

This solution would reduce the need to travel long distances, and it would improve the knowledge of local professionals about HSP (who may sit in on the specialist consultation video conference), and support for both patients and carers should allow care to be coordinated.

Jon identified that there was a burden on carers which is frequently ignored. Those who care for people with disability (not just HSP) can experience a whole range of stresses and pressures which affect their own well-being, and having carer support at these clinics would be important.

Jon has put in for funding to undertake a trial for this with three clinics in each of Devon and Cornwall, and is awaiting the outcome, but could be a useful way of providing services. To make the research funding application work, Jon will combine the clinics with similar for hereditary ataxias.

"Telerehabilitation"

Jon's final section was to ask the AGM's opinion on a potential telerehabilitation scheme. Effectively, this would be an on-line exercise programme. This has been undertaken for people with MS (Multiple Sclerosis), and Jon speculates that this could be good for people with HSP, and wants to set up a controlled trial.

Part of the management of HSP is undertaking regular stretches and exercise, and the on-line programme would give you a personalised programme to follow. Jon showed us some video

examples from the MS example. Principally, after a consultation you would be given an exercise programme to follow, which you would do, with the computer/tablet providing the timing and allowing you to give feedback.

With the MS one there was no evidence of an improvement, but there were positive subjective opinions. Jon noted that the use of the programme fell with people dropping out over the 12 week trial, and that there was limited take-up.

Jon suggested that it would be straightforward to add motivational elements and educational information as well as links to support networks (e.g. Facebook).

In discussions with the AGM, there was a general opinion that those with HSP notice the difference if they don't do their exercises, and the drop-off in use may not be so big. Suggestions were made that this could be combined with the telehealthcare, offering group exercise sessions at the clinics. Discussions went slightly off track by talking about the potential to use the accelerometers (etc) in phones/tablets to allow apps to monitor your exercise programme and provide feedback to the system.

Jon wanted to set up patient focus groups to allow the definition of this study and the firming up of the application for funding. Jon saw the trial potentially taking place with three groups in different parts of the UK so that the research would not be biased to people within a particular geographic situation.

Does physical activity improve quality of life in HSP?-Kate Winstone

Kate Winstone gave an overview of work that is currently being undertaken at Plymouth University.

Kate is examining the benefits of physical activity. It is known that there is a lower level of activity in people with neurological conditions, compared with the normal population. It is also known that physical activity can bring an improvement to quality of life.

The general question is would physical activity improve the quality of life for those with HSP? A study has shown that for people with cerebral palsy an increase in physical activity does not lead to an increase in quality of life. Kate is researching what the outcome is for people with HSP.

If her research confirms that increased physical activity does increase quality of life, then follow on work can be undertaken to establish:

- What types of physical activity? - strength or flexibility exercises, for example
- When, over the progression of HSP would this be best? - at the beginning, later on, etc.
- Which types of HSP would gain the most benefit?

Effectively, Kate's research seeks to identify if there is a correlation between physical activity and quality of life for people with HSP.

This work should have the following benefits:

- An increased understanding of the importance of physical activity
- Could lead to further research being undertaken
- An increased awareness of HSP
- Provide support for finding.

Kate observed that there was limited evidence for the general benefits of physio on people, which means that it is more difficult to obtain funding for research.

Kate then went on to describe the study being undertaken. She had been contacted by 35 members of the HSP group who wanted to take part.

The aim of the day was to assess participants using the Spastic Paraplegia Rating Scale (SPRS). Each person went to four different "stations" set up on the day testing:

- Speed of walking/stair climbing
- Muscle power and reflexes
- Memory
- Senses and sensations

After the assessment participants would have to complete an on-line survey, and after that the data needs to be analysed.

Kate will share the results of her research with the HSP group, with those involved with physiotherapy, and she hopes to produce abstracts and get a published paper from this work.

The effects of warming and cooling on HSP - Amanda Denton

Amanda reported some work that has recently been completed at Plymouth University. Many people with HSP report that walking becomes more difficult when they are cold, and Amanda reported some work which sought to investigate the effects of heat on nerve and muscle function.

The general symptoms that are reported are that people feel stiffer, that they have more falls, and that it seems to take longer for messages to get from the brain to the legs. This research was undertaken in two parts, and Amanda was principally reporting the second piece of work.

The first piece was undertaken in 2010/11 where participants were subjected to warming and cooling of their legs in the laboratory. These results showed that when legs were cooled there were negative effects, and when legs were warmed there were positive effects. This research can be seen <http://www.plymouth.ac.uk/pages/view.asp?page=38252>

This first piece of work led to a product trial of some wearable heating pads, as discussed at an HSP support group meeting in 2011. These were neoprene sleeves which could be warmed in hot water and then put on calf muscles on the front and back of the leg. These were tested being worn for periods of half an hour. The second part of the research sought to answer two questions:

- 1) Can external warming have the same effects as shown in the lab?
- 2) Is there any benefit from keeping the neoprene on?

The research sought to measure temperature, walking, tapping, blood flow, stiffness and muscle strength - all of which can be measured. Two groups were used, 21 people with HSP and a control group of 16.

The research shows that wearing the neoprene can give the same benefits as shown in the lab. Applying the warming gives a statistically significant increase in walking speed and tapping. The research also showed that there was an increase in blood flow with the neoprene, but this was also true of the control group.

As mentioned, the main part of the trials looked at the effects after applying heat with the neoprene for half an hour and some investigations were made into keeping the neoprene on. These results showed no clear benefits. The objective is to find practical application for these findings, and Amanda discussed where the research would go next, and the thought is to

undertake a case study after Christmas when it is generally cold outside. Amanda noted that people with Cerebral Palsy can report the same temperature effects as those with HSP.

HSP Research, the Historical Perspective - Dr Evan Reid

Dr Evan Reid gave a historical perspective on HSP research. He initially showed a graph with the number of research articles which cover HSP, ranging between 1947 and now, and described that research into HSP has fallen into three phases.

The first phase - the Clinical Phase, began with the original work of Strumpell and Lorrain, with Strumpell describing the condition initially, and Lorrain fleshing out the detail some more. Another researcher Dr Reid mentioned in this phase was Anita Harding who was working on the clinical definition of HSP until her death in 1995.

In this Clinical Phase, which ran through till around the 1990's definitions were made of the:

- different types of HSP - pure and complex,
- features/symptoms of HSP
- pathological anatomy
- inheritance modes
- different complex sub-types.

During the late 1980's / early 1990's the next phase started, the Genetic Phase, which ran through until the 2000's. One researcher noted was Sue (Susan) Kenwick, who was involved with the first genetic identification of HSP in 1984, which was identified as SPG1. [Various links with info/obituaries below]

In 2007 there was a leap in technology, with Next Generation Sequencing which allowed research to progress at a greater rate and at a cheaper cost than previously, now at about £5,000 per genome. [The first human genome to be sequenced cost \$100m, and costs have dropped ever since, roughly following Moores Law until the advent of next generation sequencing, with costs now at \$8k per genome <http://www.genome.gov/sequencingcosts/>].

Dr Reid said that new HSP genes were continuing to be identified, and suggested that overall there might be 100-200 HSP genes in total.

The next area where the sequencing is likely to go is in the investigation of modifier genes. There are some genes which are protective, and others which are enhancing of the action of a gene - i.e. the function of one gene can be helped or hindered by other genes.

In the background, we always have to remember what genes do. The function of a gene is to produce a protein. When a gene is mutated, like various genes are with HSP, then the end result is either that the protein is not produced at all, or there is an abnormality in the production (which may mean too much protein is produced, or not enough, or the protein is not produced properly).

This leads us on to the third phase of HSP research, the Biological and Treatment phase, which began around 2001. To describe this phase Dr Reid gave us a quick lesson in cell biology so we could understand what happens with HSP. This lesson takes the form of an analogy:

- Treat a cell as a company. It is semi-autonomous.
 - The nucleus of the cell is the head office, in charge of the company.
 - The DNA is the CEO of the company.
 - The endoplasmic reticulum (ER) and the golgi apparatus make up the manufacturing department of the company. Proteins are made by the ER and the golgi apparatus packages them up for despatch.
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- The plasma membrane, at the edge of the cell is responsible for exporting the proteins that are made.
- There are also receptors on the plasma membrane which undertake market research, identifying if more (or less) proteins need to be made.
- The power for the companies operations comes from the mitochondria.
- Within the cell there are microtubules which are like rails, allowing proteins to be moved from manufacture to export
- There are special motor proteins which are used to move things along the rails.
- The packaging of various things as they move within the cell includes endosomes, which also have a sorting function deciding if things should be disposed of, recycled or sent on.

A neuron is a special type of cell. In addition to the cell described above the neuron also has an axon, which is another method of communicating with the outside world. This is a special part of the cell which can extend away from the nucleus a distance many thousands of times the size of the rest of the cell (up to ~ 1.5m). Within the axon are microtubules which carry information to/from the nucleus. This arrangement means that the transfer of information between parts of the body is very quick, but complex machinery is needed to support this transfer. The axon from a neuron can connect to the cell of another neuron.

In the majority of types of HSP, the result is degeneration at the end the axon which then progresses. The different types of HSP affect different elements of the axon. Some affect the endoplasmic reticulum (ER), some affect the motor proteins, some the endosomes, and others some of these in combination. Spastin is a protein which regulates the microtubules within a cell. Its job is to chop them up, which helps to shape or prune them. When you have spastin HSP (SPG4) less spastin is produced, and fewer microtubules are broken up. The essential questions are:

- Why do the microtubules need to break?
- When does a reduction in microtubule breakdown cause HSP?

Perhaps the wrong receptors are on the surface of the cell, so instructions to grow/stop growing/divide/change/etc. are not received properly. This could then influence the cell's behaviour. It has been noted that with HSP BMP signalling is unregulated, but it is not known if this is the cause of HSP or an effect of HSP. If it is a cause, then drugs are available for this.

There are three areas where research in HSP is being undertaken to answer these and other HSP questions:

- Cell biology - using microscopes, stem cells, genetic edits
- Animal models - principally mice, zebra fish and fruit flies
- Human studies - HSP clinics

In summary, the genetic identification of HSP is now rapid, which improves diagnosis and allows testing. Within the cell there is an increase in the knowledge of the functions of the proteins. The proteins associated with HSP have inter-related functions. The use of animal models allows quick progression, and stem cell models are also used.

Currently there are several starting points, with potential treatment avenues identified. Thorough research is required to develop treatments.

As an example of how things are progressing, Dr Reid noted Duchenne Muscular Dystrophy. This is a genetic condition which results in muscle degeneration and death by the mid-20's. Like HSP there is no treatment. However, there has been an overall improvement as a result of the use of combined therapies. In the 1960's 80% of people with Duchenne MD had died by the age of 20. In the 1970's this had improved with 60% having died by the age of 20, and

with further improvements in the 1980's and beyond, with some people now living into their 30's.

For HSP, like the three areas of research there are three areas for treatment. The main area is in rehabilitation, where orthotics or FES may be used, or drugs given to mitigate the symptoms, e.g. baclofen. With genetic testing, advice can be given which provides clarity and frames likely progression. The other area is neurology, where assessments are made in clinic and can guide approaches for treatment.

Sportability - David Heard

David talked about Sportability, which he was one of the founding members of. Sportability was set up about 25 years ago, with the aim of providing access to sporting events to people with paralysis. This would include not only people with HSP but also people with spinal cord injury, stroke or MS.

David shared with us some stories about people who had used Sportability and gave us examples of what people had felt after taking part. The range of sports which are on offer include riding quad-bikes, archery, canoeing, sailing, scuba diving and so on. (their website <http://www.sportability.org.uk/index.php?id=3> says "archery, angling, abseiling, canoeing quad-biking, gliding, clay pigeon shooting, motor-sport and more." the list of activities also includes falconry, jet-skiing, martial arts, tennis and blokarting)

Some people who participate in the activities may have been keen participants of those sports earlier in their lives, whereas others will be trying sports for the first time. People feel a range of emotions from this, some of which are positive - for example, there is excitement and the feeling that something can be done again, and there may also be negative thoughts - for example reinforcing the perception of being disabled.

David said that one of the main outcomes from people taking part in sport were psychological benefits. Undertaking a sport either again or for the first time can begin to re-build a persons confidence and self esteem. These activities can also help to re-define someone's horizons, and people can begin to think that if they can do this, then what else can they achieve. Overall, participation in such activities can help put some purpose back into someone's life as well as getting a good shot of adrenaline as well.

David had a good selection of photos and videos of people taking part in some of these, and it was good to hear the genuine happiness and excitement of the participants. David also shared with us how different some bookings can be, in some instances people call up with a definite idea of what they want to do and are prepared to travel to take part, whereas others are not sure and David gave examples of the "negotiation process" with some people in order that they convince themselves that they really do want to take part.

Sportability aims to remove barriers to participation. So, all activities are free of charge, and there are a number of regions all over the country offering different activities. The Sportability strap line is "taking the dis out of disability". Sportability is a charity and all activities are funded by donations, so people may like to consider this as a good recipient for fundraising activities they organise or take part in.

You can find out more about Sportability here: <http://www.sportability.org.uk/>

New Members

We welcome the following new members

Alayne Horton
Luton
Region 3

Felicia Micciarelli
Wandsworth
Region 2

Lynn Hamblin
Radstock
Region 4

Michael Bradley
Shropshire
Region 7

Phil Parry
Cardiff
Region 6

Tina-Elizabeth Rudkin
Scunthorpe
Region 10

Trevor Wyatt
Thornton Heath
Region 1

Dennis Roberts
Gt Yarmouth
Region 5

Jane Jarman
Bridgnorth
Region 7

Marion Lindsell
St Albans
Region 3

Mike Perry
Gosport
Region 1

Thomas Weller
Polegate
Region 1

Tracy Askham
Skipton
Region 10

Vernon Johnson
Middlesex
Region 1

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

Useful Contacts

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