



HSP Newslink

The Newsletter of the Hereditary Spastic Paraplegia Support Group

Issue 48 – January 2020

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

It's what you CAN do that counts!



Terry Duffy and Steven Foot horse riding with the Calvert Trust at Exmoor

After a presentation by a lady from the Calvert Trust at a South West HSP meeting at Ashburton, my friend Terry and I said to each other that we must have a go at that kind of holiday, especially the horse riding that they had on offer, us being two old horsemen who had not sat on a horse for years! So we booked up for a week in September 2018. We were very lucky with the weather that week and the setting of the holiday centre on the edge

of Exmoor – it was beautiful! We were welcomed and shown to our accommodation which were ground floor en-suite rooms situated around an old farm courtyard. All the facilities are geared towards our own particular needs. We could not fault the accommodation and the food was excellent. We met the instructors who would be looking after us and were grouped with others with diverse disabilities and their carers who

were also able to join in the activities. The week was structured with two or three optional activities each day. Even though we were initially drawn to the horse riding it would have been silly not to have given everything a go.

Day one was kayaking on the large reservoir. Two kayaks were bolted together so there was no chance of us capsizing, especially with the lack of balance that comes with HSP. There was a team competition between boats with a lot of splashing about and a half time cup of tea on the water whilst we took in the stunning scenery.

Next up was the abseiling and zip wiring. Now I have no head for heights, but after a bit of encouragement and watching lesser able-bodied people go first, I asked the instructor what was the success rate and he said about 50%, I sent Terry down first then I followed. It was great fun....backwards off a rock in a wheelchair!



Terry abseiling backwards down a rock face

Day two was horse riding. They have lovely, quiet horses of varying sizes and a super indoor school with purpose built mounting blocks

where we were helped into the saddle. We were given time to get the feel of the horse and then after a walk and a trot, we went out for a ride around the centre. They also have a lovely carriage pony if you prefer a drive than a ride. Terry and I loved being around the horses again.

Next up was crate stacking. It seemed an odd thing to do but being winched up to the roof of the sports hall whilst stacking and trying to balance on milk crates is pretty good fun. Terry holds the record of 18 crates.

Day three was biking around the reservoir trail. All sorts of different types of bike, from hand pedal recumbent bikes to trikes and tandems and also Trumper mobility scooters were available to try. Again, drinks were provided and plenty of time was given to look at the scenery and wildlife.

Archery was next, again it was set up for all abilities. Terry and I got quite competitive firing arrows from our wheelchairs. There was also bush craft in the nearby woods which was a very peaceful setting around a camp fire. The only bus tucker trials were eating hot marshmallow and popcorn! They had a lovely fully accessible swimming pool and a large social area with balconies overlooking the reservoir. Although there were tv rooms, we spent the evenings in the converted barn room which had a bar and music and chatted with the other guests. One evening there was a visit from the local Exmoor zookeeper who brought in all manner of creatures which everyone could hold and touch. This was particularly good for the children.

We have recently returned from our second holiday there this September and although it rained most days, it didn't dampen the enjoyment at all.

Over all they were very enjoyable holidays which we would highly recommend to all fellow HSP'rs who are up for a challenge and a laugh

It's what you can do that counts! That's the motto of the Calvert Trust activity holidays.

Steve Foot and Terry Duffy

Chairman's Column



I hope that everyone enjoyed Christmas. With any luck this Newslink represents me catching up with telling you about things. Elsewhere you will find my reports from the AGM and from my attendance at the Spatax meeting in France in September.

At the beginning of the year people often think about making resolutions. My resolution for HSP is to make sure that I pay attention to the language which I use when talking about our experiences. Much of the language used around disability is negative, and it does not need to be that way. I want to avoid phrases like “suffers from...”, “confined to...” and so on, as these tend to portray a pessimistic view on the person you are talking about. Whilst I accept that we all have tough days from time to time I would rather portray our friendly supportive nature and demonstrate that we don't have to be limited by our disabilities.

In my last column, like at the AGM, I made a call for people to step up and help the group. There has been a limited response to this, but I still haven't heard from many at the AGM who said they would! If we cannot get more people to help then we cannot expand our friendly supportive community and reach out to support more with HSP. In reality, we need more people to help so that we can continue operating as we do.

Below, I have given an outline of some of the types of people we need to volunteer. As I said at the AGM this can be anyone. If they are keen and happy to help then it doesn't matter if they have HSP or not. These can be friends, relatives, colleagues or anyone else you know.

- **Finance:** We need our next treasurer. This would be someone who is comfortable with finance and accounting, has an orderly mind and a methodical way of thinking, has good communication skills, ensures things are followed up and is good at keeping records.
- **Meeting Organisers:** We are keen to expand our regional meetings so that we hold them in more locations closer to members. These people would be good at communicating with others, be able to organise a room to hold the meeting, network with others to have a wide range of topics discussed at meetings, and publicise the meetings that they run regularly.
- **Fundraising Coordinators:** We would like to have oversight of the various fundraising activities which go on. These people would be good at communicating with others, would help and support people who are already raising money for the group, have and publicise more ideas for ways we could raise money, and help to put those into practice.
- **Networkers:** There are plenty of opportunities to help raise awareness of HSP in wider circles, and plenty of opportunities to learn new information. This would suit people who are keen to network, don't mind travelling, and are good at keeping notes of what they hear.

I'm also keen to hear from anyone who has other ideas how they could help the Group. There are trustee roles available, you may want to help out with research, the helpline, social media, the newsletter, the AGM, group activities, and other new ideas are always welcome. A study I saw on the Australian HSP group page indicated that those with HSP often need more mental challenge to improve their quality of life: <https://hspersunite.org.au/more-challenge-needed-in-daily-life/> - helping the group could be a good way to do this.

As always, I'm always happy to hear from people with their thoughts and opinions, do drop me a line if you want to share your thoughts.

Adam Lawrence

Editor's Column

Happy new year to all members and anyone else who receives the HSP Newslink. I saw in the new year with some close friends, listening to a brilliant band down at the local pub. For me it doesn't get much better than that. However, I'm on a strict diet now because as always, I seem to have put on a few pounds over the festive period. This is mainly due to the over indulgence at this time of year, but spending more time in a wheelchair and consequently being less active certainly doesn't help to burn off the calories. This has instigated my only new years resolution, which is to do a little more physical activity on a daily basis. So far, I've done half hour stints of garden chores when the weather has allowed and on other days I've done more walking or exercises and stretches. I'll do my best to stick to this resolution.

Plans for this year's Potato Pants Festival are progressing well. I have already got three bands to agree to entertain us who've not been part of the festival before and there'll also be some of the regulars. Remember that the date for this event has changed and this year Potato Pants festival will take place on August 15th.

While I'm on the subject of music, I'm still really enjoying presenting a weekly show on local radio where I feature local music. I know a few members tune in online and if you're interested in hearing some great music on a Friday evening at 8pm, tune in to Forest FM. Apparently, it'll be available on digital radio later this year, but you can listen on line. I can't believe I've now been doing this for 18 months, scary how time flies.

Several months ago, it was actually Father's Day, my oldest daughter Jade, informed me that I'm to be a grandad. This is now imminent; the baby is due on January 25th but it's looking like it may arrive early. If it's appeared before I complete this edition of the newsletter, I'll give you further details. Jade is also getting married in May so as you can

guess, there's currently much excitement in my household.

My great friend and ex-chairman of the HSP Support Group, David Pearce, very generously left us a lot of money in his will. This of course will be put to very good use but I'm very keen to ensure that it is spent in a way that would meet David's approval. He was passionate about members meeting up, a regular attendee of the Milford meetings and he helped me get the Ashburton meetings up and running about 15 years ago. For this reason we are going to use a small amount of his legacy, to try to encourage more members to attend our AGM, so for this year's event, attendance will be free of charge.

My work as a trustee for Flying Scholarships for Disabled People keeps me very busy, especially at this time of year when we review all the applications and choose which candidates will attend RAF Cranwell for the final selection process. I notice that this year another one of our members has applied and I wish James Fitzgibbon the very best of luck. James is about the 15th HSP member to apply and most have been successful in being selected for a life changing opportunity. Applications for 2021 scholarships close on November 30th so please look into this if it sounds of interest. Further information can be found on the website: www.fsdp.co.uk

I'd like to thank Adam for all of his hard work and particularly for the excellent summaries of last years AGM presentations, which are at the end of this newsletter. This input has helped to make this an interesting edition of HSP Newslink. To maintain such levels, please keep your letters coming in. You'll notice that there's a shortage of members letters in this edition and it's impossible to produce this without your assistance.

As always, I look forward to meeting up with any of you at either Ashburton or the AGM in the coming months.

Ian Bennett

P.S. I'm delighted to tell you that I now have a lovely little grandson and mother and baby are doing well

Spatax Meeting Report, Nice, September 2019

Towards the end of September I went to the Spatax meeting in Nice, France. The meeting is held every three years, and readers may remember that I went to the previous one in 2016.

Like 2016, I found the meeting really friendly, and since 2016 I have become more involved with EuroHSP, and it was good to catch up with friends from EuroHSP and other HSP support groups. There were 159 participants from 21 countries, there were 32 presentations made and 70 posters were up. This time there seemed to be a greater proportion of papers covering Ataxia than were covering HSP. I have focussed on those papers and posters which I perceived to be more relevant for those with HSP.

Presentations

Marina Zapparoli from EuroHSP and the Italian HSP group recounted her family story, noting that with HSP there are many things that you cannot do, but that should not stop you focusing on being who **you** are, and following your dreams.

TreatHSP.net and the Alliance for Treatment in HSP and PLS

Rebecca Schüle gave an overview of the TreatHSP network. (<https://treathsp.net/>) which has goals to develop and improve treatments for HSP. They are seeking longitudinal studies and looking at recorded outcomes. There are two key issues with the network: One is that there are consent issues when combining different datasets together, and the other is that there needs to be consistency between datasets so that the whole dataset can be used together. Expert input is needed to resolve this.

IP3 Receptor Degradation – A Mutational Hotspot for Hereditary Ataxia and Motor Neuron Disease

Rebecca Schüle also presented on other HSP aspects. Genetic mutations are not currently detected for 45% of unresolved HSP cases. Rebecca also talked about the RNF170 gene. This gene has been identified with several cases of HSP, and Rebecca noted that this gene could be a "druggable pathway" - i.e. there is potential for drugs to reduce the effects of this type of HSP.

Novel Insights into AP-4-Deficiency Syndrome

Juan Bonifacino noted that SPG47, SPG50, SPG51 and SPG52 are now grouped together and called AP4. They are developing a mouse model for investigating further.

From physiopathology to preclinical trials in SPG11

Frederic Darios reported that they have developed a mouse model which replicates both the physical and cognitive effects of SPG11 (the most frequently occurring recessive form of HSP). They have also been testing a zebrafish model, and may have a cure for zebrafish with SPG11.

Dance for ataxias and spastic paraplegias

Lucy Vincent told us about click and dance <https://www.clickanddance.com/>. Undertaking new activities creates new pathways in the brain. Many people stop creating new movement paths in their brain at age 16-17, and learning to dance creates new pathways. She had everyone in the room up on their feet and was teaching a new dance step. The website has a range of different options for those who can or cant stand up, and gives a range of 3 minute videos to follow.

Exergames and physiotherapy

Matthis Synofzik reported various therapies for ataxia, but my view is that some of these approaches would equally apply to HSP. He reported that physiotherapy is an effective treatment. More interestingly, he reported that playing exergames was also good. These are games where you control the game through physical movement of a controller rather than pressing buttons on a gamepad. On the Xbox they investigated table tennis, which was shown to be good for the upper body, light race

which was good for the lower body and 20,000 leaks which was good for both upper and lower body. They also tested various games using the Wii balance board, which were shown to be more cognitively demanding. The thing to note is to change games regularly, so that you are creating new neural pathways.

Potential effects of botulinum toxin in patients with pure HSP

Alexander Geurts reported the use of Botox for people with uncomplicated HSP. They identified that if people were walking on their toes then their calf muscles were tight, and if they walked with a scissor gait then their hip adductors were tight. They undertook a programme with botox injections to relieve spasticity in selected patients. The study used a very precise way of positioning the botox injections in the places that they wanted them to achieve the benefits.

Posters

Speech Patterns in HSP

Researchers in Brazil examined speech patterns in a group of 34 patients with the most common types of HSP. They aimed to characterise dysarthria in the most prevalent forms of HSP. Analysis of those patients with SPG4 and SPG11 is presented. All with SPG11 had dysarthria (difficulty speaking). Between 20 and 60% of SPG4 patients showed differing effects. The conclusion is that mild dysarthria is present in both SPG4 and SPG11, and there is a greater impact on speech ineligibility for people with SPG11. (poster abstract 60, Characterisation of speech patterns in Hereditary Spastic Paraplegia)

Cognitive Effects of HSP

Researchers in Brazil examined cognitive changes in a group of 54 patients with various types of HSP. They aimed to characterise cognitive function of patients with pure and complicated HSP using a variety of tests. Most SPG4 (a pure HSP) had cognitive changes which are not usually associated with dementia, with effects in memory, attention and executive function. SPG5 (can be pure or complicated) scored lower in executive function and memory. SPG7 (can be pure or complicated) performed poorly on memory. The conclusion is that cognitive abnormalities are frequent in both pure and complicated forms of HSP, being more severe in complicated forms. (poster abstract 64, Are Cognitive Changes in Hereditary Spastic Paraplegia Restricted to Complicated Forms?)

EuroHSP - Mission and Goals

Information was given about EuroHSP. The group represents 10 national HSP associations from Europe. Their strategic goals are to provide a strong voice for HSP patients at a European level, to support member organisations to be more effective and sustainable, to influence the HSP research agenda so it is more focussed on patient-centric outcomes, and to secure the support of key researchers. (poster abstract 68)

Sequencing of 812 people

Researchers in Paris, France (and other places) developed a kit they used to analyse 70 HSP genes in 812 people with HSP. The kit couples targeted capture and next generation sequencing together. The technique was noted as having a higher diagnostic rate than more classical strategies. (poster abstract 76, Hereditary Spastic Paraplegia: massive sequencing of 70 genes in a large cohort of 812 cases)

Gait Timing

A study from Brazil used a range of walking tests over 25 HSP patients and 25 controls. Tests included a 6 minute walking test, a 10m walking test and a timed up and go test. On all walking tests there was a moderate to strong correlation with the SPRS scale and with disease stage, and they determined that theoretically these are sensitive enough to detect change. They plan to repeat the study after 18 months. (poster abstract 63, Natural history of movement abnormalities on hereditary spastic paraplegias: validation of timed-gait measuring functional instruments)

How HSP Affects Patient Wellbeing

I presented a poster with a summary of my last three years survey. Factors which affect wellbeing were identified in 2017 and 2018, including: pain, employment, walking ability, depression, stress, cognitive function, poor co-ordination, regular falls, and sleep. The whole poster is here: <https://hspjourney.blogspot.com/2019/11/spatax-meeting-poster-2019.html>.

Seeking More Information

Spatax have prepared their own summary of the conference here:

<https://spatax.wordpress.com/2019/10/04/summary-and-abstracts-of-the-6th-international-meeting-on-spastic-paraparesis-and-ataxia-september-20-21-2019-in-nice-france/>

You can also see the programme and the abstracts for the posters and selected papers here:

https://spatax.files.wordpress.com/2019/10/booklet_2019-version-en-ligne.pdf

Adam Lawrence

For my Mum Because she's a hero!

The HSP Support Group has been a huge help to my Mum, June Masding, who has HSP. It's offered her, and our family, opportunities to learn about the condition and to meet other people in similar situations. As a rare condition I'm pleased to have been able to talk about it and raise awareness of both what the illness is and the great work that the HSP Support Group does.



I have been running for most of my adult life and joined a local running group about a year ago, which has helped me to build my strength and stamina, and on Sunday 13th October I took part in the Birmingham Half Marathon, in support of the HSP Support

Group. It was great to take part in an organised run and gave me a goal to work towards and motivation from knowing that I was raising money for a charity very close to my heart in the process.

This was my first Half-Marathon! - I knew, with the local group that I had been training with, that I could achieve the distance, and hoped that I could do it in around 1hr 40mins.

It was very wet on the day, with heavy rains having flooded the area during the night, and at the last minute, heard that our course was to be changed!! Some of the road had been flooded and the organisers had had to cut off part of the course, meaning that our Half Marathon was now only 12 miles instead of the usual 13. Nevertheless, with the rain bucketing down throughout most of the race, I was really pleased to complete it in 1hr 22mins, and was given an estimated time of 1hr 33mins for the full Half Marathon!! I was really pleased, and was the 907th person over the line, with more than 11,000 people running.

I am delighted to have raised £410.28 for HSP Support group, and that it will be used to fund research and support those living with the condition.

A huge 'Thanks' to everyone who has kindly supported me!

Simon Masding

Spreading the word about HSP

I have been out and about a few times recently with the intention of getting HSP known and talked about by Clinicians, Researchers, NHS, Patients and carers.

This started with our membership of Genetic Alliance which is a charity focussed on gene therapies and research. I attended their AGM last year and again this year. Through them I was invited to a reception in the House of Commons where I spoke with Mary Bythell who is Head of Rare Disease Registration at NHS England and was very pleased to shortly afterwards hear Baroness Blackwood explain to people attending the reception what HSP is. Among the audience was the All Party Parliamentary Group of MPs.

Baroness Blackwood is one of the Ministers of Health. She has special responsibility for Rare Diseases, herself having a rare genetic disorder. Again she was at this year's AGM of GA though she was not there for long and I did not get to speak with her.

At the Genetics Alliance AGM, I was allowed 3 minutes, along with others, to speak, and I chose 'Consequences of Living with a Rare Disease' as my subject. There were about 100 there in a mixed audience of clinicians, researchers, patient groups and bureaucrats.

Amongst them was Lizzie ??? from NICE and we talked about a management system and expert centre for HSP. She has referred me to further reading, and I will let you know if I find anything.

I had a train home which meant I could not attend the drinks reception following the day. This highlights the need for others to help the HSP committee.

Through Genetics Alliance I attended a FINDACURE event in London. Whilst it was primarily to discuss setting registers and thoughts, I had the chance meeting with ATAXIA UK and found some link between HSP (SPG7) and Ataxia. I don't fully understand them. However my doctor in Sheffield is researching SPG7 and presented a paper to an expert society about HSP and Ataxia. His paper is on the Facebook UNITE site

Through Findacure I was allowed a 4 minute speech at Rare Disease Showcase Manchester – which just happens to be close to home!! For my topic there I chose 'Living With a Rare Disease'. In it I spoke of the 'Randomness' and 'Chance' that plays a significant part in getting the treatments that are available for symptoms and case management as collaboration and competition differ across the countries of the UK.

This was at Manchester's Bridgewater Hall where again about 100 people formed a mixed audience from across England and Wales, though mainly from the North West, the ceremonial Lord Mayor of Manchester also spoke. There were also displays and tables on the floor above for Rare Disease Groups and Researchers to meet and talk with people. By listening to others talking it became evident that there is a focus of HSP in some closed communities which may or may not have an influence of how HSP is perceived as 3 per 100,000. HSP did not have a display or table to communicate because of lack of people to talk about HSP and the Support Group.

It was interesting to see how charities smaller than ours have a high degree of influence and different approaches to publications available to members. I think HSP has the best way of communicating with members. It would be good however, to have more local support in different places in the country.

Findacure are also a charity, clearly backed by loads of money as the events I have been to are in large luxurious places. I first met them through my connections with Newcastle University where they came to talk about 'Drug Repurposing'. This has moved on in the intervening period and work is progressing apace to allow access by smaller charities.

Genetic Alliance send us frequent newsletters and our chairman has sent links to surveys by them into Care Consistency and Activity from a project being carried out by Cardiff University that he and Ian have been involved with.

Mike Cain

Fundraising News

Please read this!

Since I publicised

[Easyfundraising.org.uk](https://www.easyfundraising.org.uk) at our 2019 AGM, we now have 46 individuals raising funds for us using this facility and we have raised just over £400.

We have nearly 400 members, and no end of friends, so we should easily be able to attract many more than 39 supporters and raise significant funds for HSP with Easyfundraising. If you ever shop online and are interested in supporting the HSP charity, please register for this or get in touch. It's so easy to do. There is no catch: it really is a no brainer. For example, every time we do our supermarket shopping online with Sainsbury's, 50p goes to HSP. This alone raises £25 a year for HSP and I'm sure many of you do your supermarket shopping online. Other retailers pay us up to 3% of the value of purchase and even my eBay purchases earn the HSP Group 0.5% of the price I pay. We've just renewed an insurance policy on a car and if we'd realised that if we'd navigated to the insurers website, via the easyfundraising site, £10 would have been donated to the HSP Group.

There are thousands of retailers who support this including Amazon, Groupon and eBay, not to mention most high street stores. Use the following link and follow instructions:

<https://www.easyfundraising.org.uk/causes/hspsupportgroup1/>

As I said, any problems, give me a call but it's proving to be a great way of raising funds, we simply need to get more members and friends involved, and it really is simple.

Many thanks to the 39 people who have already signed up to this great facility.

Ian Bennett

Fundraising Activities

Over the past 12 months, many people have raised funds for us by participating in a wide range of interesting and often physical

activities. On behalf of all associated with the HSP Support Group I'd like to thank everyone who's raised money for us. You'll already have read about Simon Masding who ran the Birmingham half marathon on our behalf. Below are a few more individuals who have been fund raising for us and taking part in some gruelling activities.

Caroline Begg	Triathlon
Simon Lewis	Three Peaks
James Lane	Three Peaks
Andy Carpenter	Isle of Wight Race
Estelle Marshall	Snowdonia Challenge
Christine Shepherd	10 in 10 challenge

I'd particularly like to thank Peter Bateman and Terry Duffy for their continued hard work and generosity. They've both raised significant funds for us over many years, Terry with his wood carving sales and Peter with his pen and key ring sales.

Once again, many thanks to all who have raised funds on our behalf, all proceeds are put to very good use.

Ian Bennett

HSP Group Grants

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. The maximum grant available is currently £750. Completion of one year's membership is a requirement for applicants. If interested, please request a grant application form from the Secretary Dave Harris, details at the end of this newsletter.

Regional News

Norwich Meeting

On Saturday 12th October, the four stalwarts of HSP Norwich met over teas and biscuits. We covered a lot of subjects including complicated form filling, a little on gardening and I spoke about my cruise with my sister to

the rivers of France on the Black Watch. It's an excellent ship for people with mobility problems. The staff looked after us so well as they have done over past years. We also talked about other HSP people in our area and wished we could contact them to come to our meetings. Finally our lovely physio, Rheka, was due to come to talk to us but she was very ill. We hope when she is feeling well she will come back to our meetings. Our next one will be on Saturday 4th April. Full details are shown under 'Forthcoming Events' later in this newsletter.

Barbara Jones

Colchester Branch Meeting

We met at Feering Community Centre, near Colchester in Essex last Autumn. When we arrived, we found a children's party in full swing in the big hall and the kitchen full of food and party bags. Squeals of joy from the bouncy castle were coming through the kitchen to our hall - making conversation difficult! So, as the various nasty bugs in the area had taken their toll on our numbers this time, we hastily adjourned to the smallest room where we could shut out the sound (though, hopefully not the joy) and talk in peace, though some members fancied having a go on the bouncy castle themselves!

There were 13 of us and we enjoyed chatting our way through a range of topics including Personal Independence Payments, Personal and Carers' Budgets, symptoms, medications and exercise. One member was interested in learning more about Personal Health Budgets which was a subject which was new to most of us.

Our next meeting is on Sunday, 15th March 2020 from 2.30 pm - 5 pm at Feering Community Centre, near Colchester, Essex, CO5 9QB and, as always, everyone is very welcome to join us.

Hilary Croydon

Meetings in Cheltenham and Stockport

Cheltenham Sat 14th September

The meeting in Cheltenham was a one off because Melanie from Canada had been intending to come to Stockport but her dates did not allow. So I went to Cheltenham and also posted on Facebook if anyone wanted to join us. Four people thought they would but events overtook and prevented them.

So we had a good chat about the American, Canadian and UK approach. UK is very fortunate that meetings are held in various parts of the country at frequent intervals. The USA has a single 'conference' in a different place each year with many saving for years to go. Canada too has a distance problem. In the UK 40 miles is a long way, but across the Atlantic 600 miles is 'just down the road'.

The UK meetings are free to attend and meeting face to face is very good as people relax and speak to people they get to know.

Because of distances, the UK is privileged to have centres of excellence that are mostly accessible in a day. The same is true for most local meetings.

The sun was shining and it was warm. The restaurant was opposite the Montpelier Gardens where I spent some time.

No further meeting is planned but anyone wanting info is welcome to hold one

Stockport 21st Sept 2019

As usual we had reserved the Quaker Meeting Room as it is very accessible and we usually get about 20 people coming. However that day was very fine and sunny so few turned up. (I would have gone out for the day had I the choice)

However Mark was travelling from Edinburgh and staying overnight in his chair. Stockport is good for strengthening the arms if you go downhill first. The meeting is on one of the almost level bits of the town!!

It was really good to hear from him and showed the need for face to face meetings and chat with friends. It was sad that people

did not come but it sounds as if the meeting would have been relaxing with one not well and another holding up the ceiling!!

There was another meeting which looked very similar to ours. However we got the wheelchairs and walkers into the right room!. They also had a rare disease which I had never heard of, called 'Transverse Myelitis and seem to have similar symptoms to HSP. Will need further investigation!!

From Facebook, there is an unmet need in Scotland both for treatments and meeting places

As far as treatment and research is concerned, I am in touch with the Ann Rowling Clinic in Edinburgh to find out how they treat HSP – they treat MS through to

MND as well as Parkinsons. It's been in the news of late because JK Rowling (Harry Potter) has donated £15 million plus as a second donation.

However I am looking at a meeting in Scotland next year and suggestions would be helpful.

Thanks go to Irena for helping organise and Roham for the teas/coffees and setting and tidying up.

Next Stockport Meeting

Saturday March 14th 2020 Quaker Meeting House Cooper St

Mike Cain

Forthcoming Events

Newcastle

Saturday 29th February
1.30pm – 4.30pm
Times Square Newcastle
Actual location to be confirmed
Call Irena Pritchard: 01524 261 076 or
Mike Cain: 01614 567 531
Irena.pritchard@btinternet.com

Stockport Meeting

Saturday 14th March 2pm – 5pm
Quaker Meeting House
2 Cooper Street
Stockport, SK1 3DW
(For sat Nav use: SK1 3QL)
Call Irena Pritchard: 01524 261 076 or
Mike Cain: 01614 567 531
Irena.pritchard@btinternet.com

Colchester Meeting

Sunday, March 15th 2.30 - 5pm
Feering Community Centre
Feering
Essex
CO5 9QB
Call Hilary Croydon: 01284 728 242
tohiti@btinternet.com

Ashburton Gathering

Saturday March 21st 19th 2pm onwards
The Dartmoor Lodge Hotel
Peartree Cross
Ashburton
Newton Abbot
TQ13 7JW
Call Ian Bennett on: 01202 849 391

Norwich Meeting

Saturday 4th April 2pm – 4pm
202 Gowing Road
Hellesdon
Norwich, NR6 6PZ
Call Barbara Jones on: 01603 423 267

Milford Afternoon Tea

South/South East

Saturday April 18th 3pm – 6pm
The Clockhouse
Milford
GU8 5EZ
Call Jane Bennett on: 020 8853 4089

Birmingham Meeting

Sunday April 19th 11am – 2pm
The Kenrick Centre, Mill Farm Rd, Harborne,
Birmingham. B17 0Qx
Contact Penny Cohen on 07818 288 738
Or June Masding on 07500 584 681

Scottish Meeting

Sunday May 3rd 2pm – 5pm
Holiday Inn Express
Springkerse Business Park

Stirling FK7 7XH

Call Irena Pritchard: 01524 261 076 or
Mike Cain: 01614 567 531
Irena.pritchard@btinternet.com

HSP Support Group AGM

Saturday July 4th 10am – 4-30pm
Tally Ho Conference Centre
Perschore Road
Birmingham
B5 7RN

New Members

We welcome the following new members:

Andy Anderson Arbroath	Harris Finn Bristol	Mitesh Patel Birmingham
Angela Hutchinson Ossett	Joe Walsh Dunstable	Robert Hodge Preston

If you are interested in contacting any of the above new members, please contact the membership secretary.

2019 AGM Summaries

Rebecca Schüle

The first talk at the AGM was from Rebecca Schüle, from the University of Tübingen in Germany. We had asked Rebecca to attend the 2018 AGM, but there were various problems with flights which meant that she wasn't able to attend.

Rebecca began by giving a general description of HSP, reminding us that it is a group of diseases, all of which give rise to a spastic gait. HSP is an upper motor neuron disease - i.e. it affects the part of the nerve from the brain down into the spinal column. Upper motor neuron diseases are characterised by loss of voluntary muscle control, weakness, spasticity and clonus. There tends to be an increased stretch reflex. Where conditions affect the lower nerves the characteristics tend to be muscle atrophy, weakness, twitching and a decreased stretch reflex. HSP affects the longest motor neurons the most, so those affecting walking. After the discovery of HSP back in the 1800's, key work was undertaken by Anita Harding in the 1970's to 1990's. Anita identified that the types of HSP fell into two groups, one group with cognitive effects and one without.

Medication

Rebecca covered antispastic medication, and highlighted the need to get the right dose balance. It is difficult to get the right dose. The medication has both positive and negative effects. Positive effects include reduction in spasticity, pain, clonus and spastic jerks. Negative effects include muscle weakness, slowing down movements and reduced fine motor control and tiredness. There is a need to balance the positive benefits against the negative outcomes. Medical cannabis can have some benefits on muscles, but can also make any depression worse.

Exercise

Most people do too little exercise, and you cannot do too much exercise. In judging how much effort to put into exercise, if you take 2-3 hours to recover from the exercise that is good, but if you take 2-

3 days to recover that is not good! Gait training can be good to help with instability. It is important to find exercises which you like doing, everything gets boring after a while.

For those thinking about surgery as a treatment, it is important to remember that HSP is progressive, and this needs to be taken into account in any decisions. What is right today may not be right in 15 years time.

Bladder

Two thirds of people report bladder problems with HSP, which is severe in 5% of people and mild/moderate in about a third. There are good treatment options for bladder issues, which arise from the imbalance between the large bladder muscle against the small sphincter muscle.

Pain

Half of people with HSP report pain. Pain can arise from spasticity, arthritis, herniated discs or be neuropathic. Pain tends to increase as the gait changes and the loads on the muscles/joints changes.

Sensory Deficit

Half of people with HSP suffer from some kind of sensory deficit. This is often balance and co-ordination issues arising from changes in the sense of proprioception. Some (less than 10% get numbness or tingling. Some (30%) get ataxia which can affect coordination of upper limbs, speech and swallowing. Less than 10% get cognitive effects which are more than just memory problems, and can affect the ability to solve problems, but this aspect has not been studied thoroughly to date.

The Overall Effects of HSP

Some 10% of people start showing effects of HSP before they are 5 years old. The most frequent age for starting to show effects is 35-45. People who show HSP early tend to have a slower rate of progression than those who start to show symptoms later in life.

Half of people with HSP are still walking independently after 22 years of having HSP. After 37 years with HSP about one quarter of people use a wheelchair on a daily basis.

Those with a dominant type of HSP have a 50% chance of passing it on to the next generation, who would be affected. Those with a recessive gene have a 25% chance of an affected child if both parents carry the gene. Sometimes HSP is caused by a new mutation and neither parent has HSP.

If you know the type of HSP which you have then you are able to take these overall effects into account and know the chance of passing it on to the next generation. Overall it is expected that there are some 150-200 genes for HSP, and not all of these have been discovered yet. Many recently discovered HSP genes do not have a number. Some genes can be either dominant or recessive. The future may hold gene therapy for HSP.

European Reference Networks

HSP is covered by a European Reference Network (ERN). This network looks at how to manage HSP and has prepared management guidelines. Two main areas for treatment are drug re-purposing - i.e. finding an existing drug which can repair or compensate for a defect, and modifying the gene directly to repair the defect. No gene treatments are approved for HSP at the moment, but some are for similar conditions.

In terms of trials for all kinds of treatments there is a need to have a cohort of people with HSP who are willing to take part in the study. There is a need for national history studies to understand the population and help plan for trials.

With all treatments there is a need to be able to measure the benefit of the treatment. This could be done using a scale like the SPRS (Spastic Paraplegia Rating Scale) but could also consider a specific scale for the area being tested.

Amber Meikle-Janney

The second presentation was from Amber Meikle-Janney, who is one of the Support Group members. She gave an interesting talk about her experiences skiing.

Amber is part of the GB parasnowski development team, and she entertained us with stories about her training, her races and how she skis. Essentially, Amber sits in a rig which is attached to a single ski, and she uses her weight and arms to be able to balance and turn the ski. You can read further details on Ambers ski-ing on her blog: <http://ambermeiklejanneyskier.blogspot.com/>.

One of Ambers attributes which keeps her going is not just her excellence in skiing, but her determination to do what she wanted to do, when she wanted to do it. She has discovered an activity which she really enjoys, and she doesn't let HSP get in her way of doing this.

This approach was shown in the different videos which Amber included in her presentation, to start with Amber was seen turning and falling regularly, each time having another go. Over time it is clear that Amber has put a lot of practice in as in successive videos her skiing speed, balance and turning skills have improved, and this was further evidenced in the way which Amber described herself in each video, showing her confidence and her enjoyment from skiing.

This positive approach to life stands Amber in great stead, and she uses this approach throughout her activities. She concluded her presentation with a few examples of the other things which she gets up to, including developing her wheelchair skills so that she is able to get into as many places as she can and do what she wants to do when she gets there. She has also taken part in a range of outdoor experiences including boats, camp fires and hand cycles. She was also very pleased to show us a video of her latest wheelchair attachment - a motor, allowing her to effortlessly whizz along the road.

Amber is a great example of someone using a positive mindset to achieve what they want to do and having fun along the way. Excellent!

Brandon Scott-Omenka

The third presentation was from Brandon Scott-Omenka of the Carers Trust. Brandon explained that the Carers Trust is arranged into different areas, and therefore the services which are offered by the Carers trust in one area might be different from the services in a different area.

Brandon outlined that carers are people who provide care for another. Generally this would include adults caring for parents, relatives or siblings, but can also include children caring for adults or siblings, and parents caring for children. Carers can be any age, Brandon had examples of people aged between 6 and 96.

Caring may include:

- Having an emotional burden of worry about the person being cared for
- Needing to physically assist the person with their mobility
- Spending time on caring activities, shopping etc.
- Having relevant knowledge, medical and other, knowing medications, procedures etc.
- Organising and attending medical/professional/health appointments etc.
- Being and advocate for the person - assessments, health plans etc.

Carers can often find caring for another stressful, and many feel unable to cope. Often there are role reversals, and family relationships can breakdown. Sometimes children feel isolated or are bullied.

When seeking help there are also pressures on the healthcare system meaning that help may be delayed, restricted or not available. Sometimes the person needs to end up sooner in residential care.

Being a carer can be lonely and isolating. New carers are often daunted or overwhelmed by the responsibility of providing the care. Carers also have to balance their own health, with some being exhausted, suffering from injuries or having their own health problems.

With support, these burdens on the carer can be reduced. The carers trust aim to provide support to help carers. This includes:

- Emotional support - a listening ear
- Befriending - linking carers with others
- Carers assessments - helping to get access to support
- Support group access and face to face support
- Events and activities to give a break from caring
- Providing information and advice
- Training and workshops
- Advice on future and contingency planning
- Young carers clubs

Brandon said that there are often tears the first time a carer visits as it is the first time they are being asked how they are. This is a link to Carers trust website: <https://carers.org/> where you can find out more information.

The relevant legislation if you are in England is the Care Act 2014, with other legislation in other parts of the UK. (<https://www.carersuk.org/help-and-advice/practical-support/getting-care-and-support/care-act-faq>). Under the Act carers can ask for an assessment of their needs which runs in parallel with an assessment of the needs of the person being cared for. Decisions on planning about care should take the carers wellbeing into account.

Brandon offered two statistics - the care provided by friends and family was worth £119 billion in 2011, and the 6.8million people who provide unpaid care saved the state £132 billion in 2015.

Dr Mohammed Belal

The final talk of the day was Dr Mohammed Belal of University Hospital Birmingham who talked on bladder management with HSP.

He began by giving some basic information. The bladder is a low pressure reservoir for fluids, it is a void within the body. The bladder is controlled from the brain with nerves between the bladder and the brain. The average person urinates 7 times in a 24 hour period, with young people urinating fewer times than older people.

An over-active bladder is one which gives the person urgency to go to the toilet before the bladder is full. The bladder is contracting without the permission of the person. In the normal population some 15-20% have an overactive bladder.

Dr Belal described two areas of interest in reducing bladder effects.

Lifestyle Changes

Have your last drink of the day 1-2hours before bed. Even taking a sip of drink during the night will create more urine. As you get older more urine is generated at night.

If you snore then more urine is produced, so getting a better pillow to reduce snoring should reduce the amount of urine generated.

If your wee/urine is clear then you are drinking too much. One litre of fluids per day is enough.

Tea, coffee and alcohol are diuretics whose job it is to generate more urine. Reducing intake of these drinks can reduce the amount of urine.

Bladder training can be effective. Even trying to hold back another 10 minutes can be beneficial. It is wise to keep a bladder diary to identify how often you go. Exercising the pelvic floor muscles can help. Emptying the bladder before going out and before going to bed can help.

Reducing weight and stopping smoking can also help.

The body takes a couple of weeks to adapt to any changes, so perseverance is needed when investigating these factors.

Medication

The aim is to reduce pressure in the bladder. This can be achieved in two ways, self catheterisation or using medication

Antimuscarinic drugs reduce bladder spasms. These come in a lot of different forms with different symptoms and risks. treatment can be limited by side effects including a dry mouth, constipation and effects on the central nervous system.

Botox injections can increase bladder capacity.

Infections are common when starting to use self-catheterisation, which usually need a course of anti-biotics to treat. It is necessary to practice.

It is possible to combine more than one medication.

Dr Belal also described a study which retrospectively looked at 33 people with HSP and their urological function. (link to abstract: <https://www.ncbi.nlm.nih.gov/pubmed/30848841>).

The most frequent symptom was urgency, most use medication, one third use self catheterisation and 10% have botox injections.

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