HSP Newslink

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

Issue 30 – September 2013

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

Extra Reward for Extra Effort

Mark Tomlinson with the George Stewart Memorial Trophy at the FSDP presentation ceremony at RAF Fairford in July

Each year Flying Scholarships for Disabled People (FSDP) awards a special trophy, the George Stewart Memorial Trust Award, to recognise one scholar's particular efforts. Every scholar works hard during the flight training process and the benefits soon reveal themselves in each one.

Several years ago the FSDP scholar selection process took place at Biggin Hill Airport. George Stewart was an air traffic controller at Biggin Hill at the time, and was greatly impressed by the applicants and the aims of the Charity. He left FSDP a small legacy in his will and the George Stewart Memorial Trust award was born.

However, it's often obvious that someone in particular has come that bit further in their personal journey. During the training

Trustees visit the scholars at their various flying schools and sometimes the positive change in an individual is immediately obvious, which is additionally backed by their instructor's observations.

Mark Tomlinson is one such student. He told us that **prior to learning to fly, he felt invisible especially at work amongst his colleagues and bosses.** Flying endows a pilot with a very special form of confidence, most adults can drive a car, but not many can fly! His determination to overcome his fears, from his HSP condition and the way it impinges on his everyday life, to finding that initial special courage to take to the air did not go unnoticed.

And in recognition of Mark's enormous mental as well as physical efforts, he was awarded the George Stewart Memorial Trust award. **Mark is not longer invisible!**

Susie Dunbar FSDP Trustee

Chairman's Column

I seem to regularly begin my column with an apology for the delay in producing each particular issue of our newsletter. On this occasion I'm going to use school holidays, family visitors and a gorgeous summer as my feeble excuse. Just as I was about to put pen to paper, along came the Bournemouth Air Festival where I thoroughly enjoyed three consecutive days in the sunshine watching amazing air displays highlighted by the Red Arrows and the Battle of Britain Memorial flight. All of this was at no cost. The disabled parking facilities are very good and for anyone who hasn't been I would strongly recommend this event. I don't believe there is a better venue for an air show in the UK.

Our AGM back in June now seems a fair time ago but as always it was brilliant seeing so many of our members enjoying the day. The minutes of the AGM can be found in this Newslink as can a summary of all four presentations. I'd like to thank Adam Lawrence for providing us with such accurate summaries of these presentations. Physio Function also attended the AGM and offered free assessments where members had the opportunity to try different FES systems.

On the subject of the AGM, nobody came forward to offer help or was nominated to join the committee. One of the main purposes of the AGM is to structure the committee for the year ahead and we currently seem to be very stagnant at committee level. I believe that for any organisation to move forward, new blood is needed from time to time. I for one could certainly do with some assistance as I don't have the stamina I had when I first got involved 10 years ago. If any of you would be interested in helping out the committee would be very keen to hear from you.

I'm looking forward to the next meeting in Ashburton, Devon on October 26th. Professor Jon Marsden will be joining us to update us on the work he is doing at Plymouth University regarding the effects of hot and cold temperatures on individuals with HSP. I'll be emailing invites to all members in the South West nearer the time.

Another meeting for our members in the North West has been organised on November 16th. We've also organised a meeting in Norwich on October 19th for members in Region 5 to get together. Details of both can be found on page 8.

Later in this issue you'll read about Peter Bateman's Ukulele concert in Porthcawl. I hear that Peter is a bit of a star on the ukulele and I thank him for organising this fun event and raising over £300 for the Group.

Further thanks to everyone else who has raised funds for the HSP Support Group over recent months. To name just a couple, Leanne Piccirillo jumped out of an aeroplane and raised in excess of £100 and the Enfield Grammar School Lodge have very generously donated £750.

I visit our Facebook group (Hereditary Spastic Paraplegia's Unite) regularly and I notice that it now has more members than our Support Group. It's certainly a great resource for sharing information and seeking advice. If you haven't yet discovered it, please sign up and make the most of all it has to offer.

Please keep the newsletter material rolling in and I look forward to catching up with many of you at forthcoming meetings and events.

Ian Bennett

The search for greater knowledge and more effective drug treatments for HSP

In the neurogenetics clinic in London we diagnose and manage the treatment of many patients with HSP. Regardless of the genetic cause, one of the commonest questions asked is why are there not more effective drug treatments for HSP that stop progression or reverse this disorder? This is quickly followed by how rapidly will the disorder advance and what will the future hold?

A major bottleneck in the improved understanding, discovery of promising treatments and biomarkers of HSP progression is the development of a detailed patient registry and blood sample biobank. A registry would allow patients to be grouped by the gene that causes their HSP and therefore, provide a greater understanding of how HSP progresses and attract research funding and pharmaceutical companies with specific therapies to investigate. The registry of HSP patients that are willing to be contacted and informed about trials to see if they are interested in participating will be essential for future UK drug trials.

A second, and equally important question is how quickly will each genetic type progress and what disabilities will patients develop. This is known for some genes but little is known for others. In addition, as part of the registry we would have all blood results and MRI scans to follow progression and also blood samples as part of a biobank that would allow biomarkers to be tested by researchers and pharmaceutical companies. A further benefit to patients of the registry is to make sure they have had the correct investigations carried out.

The development of a registry and biobank is an important task and will depend greatly on the help from patients and the HSP society.

The HSP registry and biobank would consist of:

1. Details of how each patient's HSP started and progressed with a yearly rating scale.
2. MRI, other scan imaging results, along with the results of other tests
3. Results of genetic tests and detailed family tree
4. A blood sample from each patient for DNA, serum and cells. This would be done for an accurate diagnosis (particularly important) and every three years to develop markers of progression.

The registry would be administered at the National Hospital for Neurology and Neurosurgery and we would aim to enrol as many patients in the UK as possible. Patients would have a unique number and collaborators and other researchers would only be able to see this number, whereas we would have full details to enable contact and follow up. The UK HSP registry would also be important for collaboration with other HSP societies such as the European SPATAX organisation.

Ideally we would enrol everyone in the UK with HSP in the registry and in the next few months we will be trying to spread the word as I think the registry will be of great benefit to HSP patients and the development of treatments and markers of HSP progression.

Henry Houlden

Professor of Neurology and Neurogenetics, The National Hospital for Neurology and Neurosurgery, Box 12, Queen Square, London, WC1N 3BG, UK.

Email: hhoulden@ucl.ac.uk

Useful Information

Sportability

Using sport and other challenging pursuits, Sportability seeks to put the purpose back into lives shattered by paralysis; to get the adrenalin rush back into a lifestyle coralled by a wheelchair or confined by crutches; to take the 'dis' out of disability and focus on ability. And to inspire the thoughts " If I can do this what else can I achieve ? There is life after paralysis."

What It Is!

"I'm not a sports fanatic or anything, but proving to myself just what I'm capable of has helped in every facet of daily living. There really is no better way to build a life worth living after paralysis. I can tell you that from the heart."

That quote sums up Sportability. The Charity has one aim – to provide **sport and challenging pursuits for people with paralysis.**

It's not about creating elite sportsmen and women. It is about ordinary people getting off their crutches or out of their wheelchairs and enjoying the fun, the challenge, the rush, the social chit-chat, the whole Sportability Experience and putting the buzz back into life!

Starting modestly in 1989 and we are now active in 13 regions around the UK, and still growing! Young, old, male, female, makes no difference – there's a sport or activity out there that can fire your imagination, get your pulse racing, lift your spirits high and put a smile on your face.

Activities include archery, sand yachting, quad biking, microliting, gliding, sailing, clay pigeon shooting, kayaking and more! Visit our website for further information.

Just Do It

If you, or someone you know, would like to participate in our activity programme, then please feel free contact us.

And talking of "free" that's the cost – our events are available free of charge. We don't want any barriers to participation.

Why do we do it? If we can help just one person, perhaps someone currently lying in a hospital bed thinking life has to come to an end, to turn back on to a life worth living it's job done !

Full details can be found online at:

www.sportability.org.uk

email: info@sportability.org.uk

Sportability is dedicated to re-building those lives shattered by paralysis.

HSP Group Funds

The HSP Support Group is now in a position to support research projects with a maximum grant of £10 000. Applications for such support are welcome. Please apply in writing.

Funds are also 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.

Survey for People with HSP

Readers may remember my article a few editions ago about my HSP blog. For a new phase in the blog I've set up a survey to capture information about people with HSP. If this is successful I'll repeat each year with a different topic. I would be pleased if you'd consider spending a few minutes to answer my questions. I aim to report survey findings on my blog in the New Year.

http://www.surveygizmo.com/s3/1360676/HSP-Symptoms-and-Mis-diagnoses

Adam Lawrence

Fergus Palmer goes "Through the Keyhole"

Turning 70 years of age can be an eventful time in ones life. One can look back and reminisce. In my case, I reflect on my condition. HSP was diagnosed quite late in my life. I often think of that television programme, "Through the Keyhole" the clues are there; as we go through the keyhole. Going through the keyhole of my life, the clues were there!!

Three months before my 17th birthday the family sent me on an outbound course to Aberdovey, Wales, in November 1959 for a holiday. I was very gullible in those days! Holiday it was not!! Character building it was! In the last week, when pushing my canoe, I stumbled, was duly knocked unconscious and was rushed to sick bay by motor launch. I returned home with severe back pain. My sister, Judy claimed the wimp returns!!

United States

In March 1962, I emigrated to the United States of America, and in June 1964, I was duly drafted into the US Army. I could write a book about my experiences. After all the tests of the day, I was diagnosed as suffering from dyslexia, which could explain my poor school results. The drill sergeant claimed, no one could march behind me with my gait. Why did I not join the Royal Navy and not his Army? I was then posted to Fort Knox Kentucky, to become a cavalryman i.e. tanks. This meant I no longer marched. I was then posted to South Korea, spending 13 months patrolling the Korean Demilitarised Zone.

Back to England

During the next 25 years my condition deteriorated, but not to the extent I could not work. I returned to England in 1976. My gait to all concerned was abnormal. My brother, Hamish, said "You walk like a blob of jelly". It was not until I met my wife, Anne, at the age of 50 who suggested I should visit my doctor, who in turn thought I could be suffering from cerebral palsy. The neurologist diagnosed FSP, and wondered why this had not been picked up when I attended Outward Bound. The US army he claimed would take any one!!

The next 10 years I basically spent in denial. I drove an average of 800 miles a week as a dry cleaning technician. Most of my customers thought I suffered from arthritis, until one day my collapsible walking stick, which I folded up before entering a shop, sprung out of my wax jacket and frightened the customers, as well as me. The manageress said, "Don't worry that's just Fergus, pretending he does not need a walking stick". I learned something that day, whom am I kidding. Be Yourself!!

There is Help Out There

I can remember our first meeting with Ian Bennett, Region 4, it was in an hotel outside Exeter. Ian suggested I contact Odstock in Salisbury and try the Functional Electrical Stimulation FES. I did, and this week made my tenth annual visit. It is a godsend for me. The FSP Information Booklet is most helpful. I also use the Neuro 4 Trophic Stimulator, issued by the Lindens Clinic. This helps to strengthen the quad muscles and was introduced to me by a neuro-physiotherapist. May not be everyone's cup of tea. Hydrotherapy is also of great help

Conclusion

I consider myself very lucky. I have made many disabled friends over the past years and they all have one thing in common, it is called a positive attitude. I must admit, some days I say "Thank you God, but why me" Then I remember attitude, have I got it right today?

Fergus Palmer Region 4

Members' Letters

Dear All,

I remember writing early in 2011 how I thought we should try whenever possible to seek rich experience, as the indelible memory of it can well be such a fillip when life's otherwise proving irksome; further, that, when pushing the boat that bit further out, we are likely to need some assurance that we are not being simply reckless, that peace of mind that is beyond our comprehension and God-given.

Since then, having steadily become more dextrous with my elbow crutches, I have, with that peace that puzzles a good few people, ventured into unusual more challenging environments and safely returned marvelling at so much I've seen, more than likely once in my lifetime. Having for a number of years wanted first-hand three-dimensional experience of the Northern Lights, I went earlier this year to arctic Norway, armed with optional spikes over the ferrules of my crutches, only then to totally fail my ambition! They are ellusive, after all, but the impression of them that I bought in Svolvaer from a local photographer and the awe-inspiring spectacles of brilliant sunshine playing on the snowclad environment admirably compensated. I took a tumble or two, but no lasting damage! The last night was spent quite extraordinarily 'simmering' at minus 5°C in the Igloo ice hotel, where it is possible, oddly enough, to sleep complete with wall-to-wall ice around you.

Let alone the personal fillip that I certainly got from all this, any notion among those unafflicted that our disability may render us 'less alive' is most certainly dispelled by that sort of revelation and the lively conversation that usually follows. While there can be no denying our weakness, the strength left in our upper bodies unaffected by the paraplegia, can, I've found, gradually be made to achieve so much more, tethered only, I hope, by our Maker.

John Moule

Della's thoughts on Fatigue

Fatigue can be an all consuming condition. It's not visible to others, except, maybe the drawn look and dark shadows under one's eyes. Many, who do not understand, think the afflicted person is being lazy! This is not so and can be very distressing for the individual, being accused of not bothering or being selfish.

Two states of fatigue that I have witnessed

1 - Depression Fatigue

I have, in the past, suffered from Depression fatigue. It's an all consuming feeling of wanting to hide away. To not face up to realities of life. Doing the minimum daily chores, then back to bed. Same with work. Go to work. Do the necessary, then home. Because, so exhausted with the worry of getting from A-B, whilst at work or home. Making the effort to put one foot in front of the other. Worrying that people were thinking bad things about you. The feeling that life did not have a place for you. It's more an insular thing than physical. The only way I could get out of it, was concentrating on something positive. I did not know it at the time, because I was concentrating on my illness. HSP etc. People were trying to tell me this, but I could not see it. A case of being at the bottom of the pit and not trying to get myself out. It is the individual that has to, with help, decide to change their outlook on life. Having others to care for you, whilst in this pit, is essential. It is hard for them also, because when we are like this, we tend to push the one's that love us the most, away.

Yes. I have been back there a few times since then, because of major upheavals in my life. But, nowhere near the first time. It's more like a typical reaction to events in life. Necessary to go through, to move on in life.

2 - Extreme Fatigue

I have and do suffer from Extreme fatigue. Fatigue is a big part of my life, due to HSP. Do not get me wrong! I am, in no way depressed, mentally. It's the extreme effort of trying to do normal activities in everyday life. It takes so much more energy to do simple things, that used to be effortless. I used to do the housework, go to work, do the gardening and shopping in a day. Now it takes most of one day, to do anyone of those listed. I have had to slow down immensely as HSP has progressed. This, at times, has made me drop into (Depression fatigue) at certain points. Mainly, when so fatigued, that one feels useless. I forget, when feeling this way, that it will soon pass. I am getting used to it now. I know, to rest when I can't physically do life activities, without an extreme effort. It's a case of having to just leave stuff and rest. Be it sleeping, or just resting and doing the minimum. It is totally different from depression fatigue. it's mostly physical, total drainage of energy. Legs feel heavy. Balance is way off. Falling around, stopping my fall on furniture or walls. I now have a tray trolley walker, to stop me falling and hurting myself, also, a walker in the garden. When extreme fatigue hits, I cannot think properly or talk without slurring. It is hard to concentrate, when someone is talking, taking in information, let alone remembering anything more than a few minutes. Clonus is more noticeable when like this. It feels like someone has turned on a tap or zapped all the stuffing out of my body. This, I am sure, could be described as being drunk. The symptoms are similar, or the same. I can assure you, no alcohol is consumed at this time! I call it "Drunk with fatigue".

In my opinion, I feel that Extreme fatigue can produce Depression fatigue. As, when Extreme fatigue hits, we feel useless and life is oh so difficult. If you are not in the right frame of mind, it can fool your mind that things will not get back to "normal". As discussed recently with a fellow HSP'er. I now know my body and mind. That Extreme fatigue will not last. I enjoy myself when I can, knowing full well that Fatigue will hit. So, I do, when I can and don't, when I can't. I'm not saying that I'm happy with it. I hate it, when it is bad. But, I DO know it will not last. It's a case of listening to one's body.

A positive outlook is essential to enjoying life, despite life's lows. Always good to have things to look forward to and friends and family to talk through any worries in life. I am in a happy place. Yes, life could be better, but that is the same with most lives. Could be a lot worse.

I hope I have not DEPRESSED anyone, with my thoughts? Sorry! Be nice to hear others thoughts on this subject. I know, everyone see's things differently.

Della Brookman Region 3

Regional News

Region 6

Ukulele Concert

Held on 8th June 2013 at the Grand Pavillion in Porthcawl, South Wales.

The Porthcawl Ukulele Band were in concert with our very own Peter Bateman in the line up. What a great evening was had by all who attended, enjoying lots of singing and great entertainment. The room was full of lots of HSP Members and friends and guests to support us. It was fantastic! At the interval ice creams and drinks from the bar were available. In the second half there was lots of audience participation. After the concert we sat outside the Pavillion to enjoy a drink and catch up in the lovely summer sunshine. Well done Peter.

Angela Barnicoat Region 6

Forthcoming Events:

Afternoon tea Regions 1 & 2

Sunday November 17th 3pm – 6pm
The Clockhouse Milford
Call Jane Bennett on: 020 8853 4089

Region 3 Meeting

August 16th 2014 1.30pm – 4.30pm
The Orange Tree Public House
100 Stevenage Rd, Hitchin, SG4 9DR
Contact Della Brookman: 07710637941
or Ian Kitchen: 07540476735

Region 4 get together

Saturday October 26th 2pm onwards
The Dartmoor Lodge Hotel, Ashburton
Call Ian Bennett on: 01202 849391

Region 5 Meeting

October 19th 1-30pm – 5-30pm
Premier Inn, Prince of Wales Road,
Norwich,NR1 1DX
Call Ian Bennett on: 01202 849391

Region 8 Meeting

February 15th 2014 1pm
The Aviator Hotel, Sywell Aerodrome,
Sywell, Northamptonshire, NN6 0BN
Contact Leanne Piccirillo: 07825516368

Region 9 Meeting (North West)

Saturday November 16th 2pm – 5-30pm
St. Helens South Premier Inn
Eurolink, Lea Green, WA9 4TT
Call Ian Bennett on: 01202 849391

New Members

We welcome the following new members

Angela Jones
Hertfordshire
Region 3

Brian Sanderson
Accrington
Region 9

Frances Ibbett
Bedford
Region 3

Ian Winton
Wirral
Region 9

Norbert Faulkner
Fleet
Region 1

Sarah Jelliman
Cambridge
Region 5

Trever Dyer
Truro
Region 4

Brian Hubbard
Bedfordshire
Region 3

Claire Blackburn
Kent
Region 1

Hilary Croydon
Bury St Edmunds
Region 5

Lorraine Ibbett
Northamptonshire
Region 8

Sandra Parr
Islington
Region 1

Sarah Rannard
Wirral
Region 9

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

HSP Support Group 2013 AGM Minutes

Saturday 15th June

The Trident Technology and Business Park, Poseidon Way, Warwick

Apologies for absence were received from Simon Hubbard, David Pearce, Jane and Michael Bennett, Michael Horsman & Andy Barrett.

The Minutes of the 2012 AGM were agreed to be a true record: proposed for acceptance by John Moore and seconded by John Mason.

Committee Reports:

a) By Chairman, Ian Bennett. Thanks were given for all of the local Meetings, we held a meeting in the North West at St Helens and we hoped that this would become a regular event. Ian reminded us that membership any local area could hold their own meetings at any time. He then added the strong reminder that anyone who needs mobility aids or equipment should propose to the Committee via Secretary David Harris to seek a Grant. Ian pointed out that, for the first time, our overall membership had dropped (this year 332 as against last year's 344), but this was in part due to the fact that Treasurer John Mason had issued some "last reminders" to people who had not responded to payment-chasers over recent years. However our Honorary Members have increased from 48 to 53. Gratitude was again given for members who have subscribed under the Gift Aid Scheme. Ian concluded with the comment that any application to join the Committee was always welcome.

b) Secretary David Harris acknowledged and reinforced all of Ian's points.

c) The Treasurer's Report by John Mason had been handed around to all attendees: very clearly produced and summarised by John himself, the Report was accepted unanimously. Its accuracy was proposed by Penny Cohen and seconded by Andre Wheeler. Thanks were expressed to Sonya Mason for her assistance through the year and to Theo White as Auditor.

Election of Officers:

Chairman, Ian Bennett was proposed by Maggie Gillson and seconded by Della Brookman.

Secretary, Dave Harris was proposed by John Mason and seconded by Roy Myers.

Treasurer, John Mason was proposed by Lorraine Saupé and seconded by Richard Williams.

Helpline, Stephanie Flower was proposed by Carol Moore and seconded by Des Williams.

Executive Committee Member Simon Hubbard was proposed by John Mason and seconded by John Moore.

Second Executive Committee Member John Moore was proposed by Terry Duffy and seconded by Della Brookman.

All the above appointments were approved by the meeting.

Any Other Business No items were raised.

Speakers:

The AGM was followed by excellent addresses from 4 of our professional supporters:

Liz Redmond, Genetics Nurse Specialist at the National Hospital of Neurology, focusing particularly on stress and anxiety management, and peoples' personal means of coping with it.

Professor Henry Houlden from the National Hospital of Neurology and Neurosurgery. He handed out a HSP Questionnaire for completion and return by those willing to assist. His talk, and the Questionnaire, focused clearly on the strategy of diagnosis and on the widespread range of associated symptoms.

Dr. Siva Nair, Consultant Neurologist at the Royal Hallamshire Hospital informed us of the forthcoming proposed research looking at comparing the use of FES and splints for individuals with HSP.

Alison Clarke, Clinical Specialist Physiotherapist gave her expert views on how people cope with the variety of assistance and treatment given, including walking aids. This was excellently put forward and well understood.

A Summary of the four AGM Presentations

Looking After Yourself (Liz Redmond)

Liz Redmond is a neurogenetics nurse at the National Hospital and gave the first presentation of the day titled: Looking After Yourself. Her presentation discussed trying to maintain a positive mental wellbeing.

Changes in mood can end up in a vicious circle. A low mood can give rise to poor motivation. Poor motivation can give rise to low self esteem. Low self esteem can give rise to low mood. 80% of people with chronic disease suffer from low mood at some point. Symptoms of low mood can include fatigue (being tired, lack of energy etc.) and anhedonia (a lack of interest in something you would normally be interested in). Feeling low for a few days may be OK but Liz advised that if you're finding yourself low for a period of weeks then its time to seek help, your GP or a specialist.

To look after your mental health you need to be mentally active. Things which you can do include:

- Make an effort to plan your time
- Plan a treat into your day
- Make time to spend with friends/family
- Recognise situations that upset you (and have strategies to deal with these).

One technique which you can use is CBT (cognitive behavioural therapy) which helps with identifying actions/strategies for dealing with these situations which upset you. Essentially it seeks to identify what it is that upsets you, to then look at that issue from other alternatives, and then to prepare appropriate responses to help you regain control of the situation.

Liz mentioned some web-based CBT software:

Mood Gym: https://moodgym.anu.edu.au/welcome

Beating the Blues: http://www.beatingtheblues.co.uk/ (Post AGM note: £150 fee for this)

There are also other ways which you can raise your mood:

Exercise - Exercise increases the levels of serotonin in the body. Undertaking exercise gives you time to monitor your thoughts, can help you relax and be part of a daily structure. For those with HSP stretching is a good exercise to try, and Liz suggested focussing on the muscles that you are using in stretches.

Positive Role Models - Thinking about someone who inspires you (whether famous or not) can give you an alternative viewpoint. You could consider what you can learn from them, how do you think they would behave in the situation, how could you do things differently to be more like them (etc.)

Social Life - Having a social life is very important. You should make time to spend with friends. People with low mood tend to reduce their social circle. If you end up like this for a while then your friends get used to you not being around and it is harder to get back to where you were. Liz advised to fight hard against this and make time. It is important to have friends around you. One step further would be to make new friends or try a new hobby.

Setting Goals - setting yourself a goal can be good, but they must be "Smart". I'll use weight loss as an example:

- Specific - i.e. "I will lose weight" rather than "I will be more healthy"
- Measurable - i.e. "I will lose 1 stone" rather than "I will lose some weight"
- Attainable - this is about setting a realistic target (which in this example would depend how overweight you were to start with)
- Relevant - make sure the goal you set is worthwhile
- Time-bound - You to set a realistic timeframe over which you plan to do this.

You can read up on smart objectives elsewhere (e.g. http://en.wikipedia.org/wiki/SMART_criteria) If your target is to, for example, lose 5 stone in weight then it would probably be better to set yourself a separate goal for each stone of weight, and there is nothing wrong at all with reviewing and adjusting your goal as you progress.

Complimentary medicine - massage, aromatherapy, acupuncture, pressure point therapy (etc.) can all make you feel better about yourself, and for those of us with HSP there may also be some muscle relief as well.

Liz's presentation concluded with an overview of fatigue - factors which can affect fatigue include: changes in sleep patterns, medication, mood, exercise. If you find that you suffer from fatigue then it becomes important to plan and to conserve energy - so plan to have a rest if you need to, prioritise your activities accordingly, organise your work/home so that it is easier to do things and check your posture.

Those with HSP could try looking at the expert patient programme: http://www.expertpatients.co.uk/ which provides tools and techniques to help people manage long term health conditions.

Getting the Correct Diagnosis (Prof Henry Houlden)

Prof. Henry Houlden works at the National Hospital and gave an overview of HSP and the 'typical' case which he and his colleagues see at the hospital. He then went on to discuss various treatments and some current research, and finished with some observations about drugs.

There are two types of HSP - Pure and Complex. With pure HSP the three main areas to cover are legs, bladder and back pain. Requests for amputation of the legs is not unknown and most

patients have some bladder issues. Bowel issues are very common as well as bladder issues. With complex HSP a range of other issues also arise including Ataxia (affecting the balance), memory, seizures and deafness. HSP is caused by an error in the genes.

Generally HSP is passed down from the parents although occasionally HSP arises without any family history, which is called a "de novo" gene mutation. There are three different inheritance patterns - Dominant, where the presence of the mutation gives rise to the condition (most commonly SPG4, SPG3A and SPG31), Recessive, where the mutation is needed in both parents to give rise to symptoms (most commonly SPG11) and the rarest X-linked inheritance. If you know which type of HSP you have, you can predict potential problems in the future.

People have varying reasons for choosing to have a genetic test following a clinical diagnosis, and there are pro's and con's. Having the test can confirm the diagnosis, and can inform treatment, aid new research and examine the risk to other family members. The current cost of a genetic test is about £500.

The typical patient seen in the Neurogenetics Clinic (on Friday afternoons) had some onset in their 20's, usually tripping or scuffing. When they look back they realise they had some difficulties in sports at school, they may have some weakness due to the stiffness and it has taken some 10-12 years to end up with the correct diagnosis.

Prof Houlden said that he would rather patients with HSP came to visit him wearing old shoes rather than new, so that he can see how much and where they are worn

Treatments include: Physio on the legs and orthotics, Prescription of Baclofen, Self catheterisation, The use of high walking sticks, new hips and knees. Prof Houlden covered each briefly (except Physio, covered later in the day).

Baclofen can make you tired, and there are alternative medicines which you could use, but each has its side effects.

If there are bladder issues, then the first step is to treat underlying problems first, e.g. prostate. Bladder problems with HSP will not go away. There is also medication that can be prescribed to help, including Detrusitol, again with side effects.

The use of high sticks, like Norwegian walking poles, can be a help because they keep the body more erect and they open the body up. The use of walking aids was discussed, and the view is that using walking aids is not the start of the "slippery slope" towards a wheelchair. The majority of patients who use walking aids wished they started using them earlier. The patients who progress the best are those who keep themselves active, using their aids and get out and about. The patients who progress the worst are those who sit at home all day and do nothing.

Having replacement joints is an option, and the suggestion is that hips would be replaced before knees, there being a longer rehabilitation period for knee replacement for patients with HSP.

Prof Houlden had recently been to the International 2013 conference on spinocerebellar degenerations at the European SPATAX (http://spatax.wordpress.com/) where there are groups of researchers looking for HSP patients for trials.

There are no drugs that can reverse or halt the condition, but there's some movement on stem cell research, trying to reprogram stem cells into neurons, and then getting these to go to the affected cells. The reprogramming is possible, but no-one knows how to make them go to the affected cells.

Cannabis would be a helpful drug for HSP, as would Sativex (a cannabinoid medicine for the treatment of spasticity due to multiple sclerosis), but this is not licensed for HSP. A discussion

ensued about Botox, with some members of the audience finding it useful. Some patients benefit from a Baclofen pump. HSP is perfect for FES because all the nerves in the legs are intact.

Research Update (Dr Siva Nair)

The third paper was a research update on treatment, given by Dr Krishnan Padmakumari Sivaraman Nair of the Royal Hallamshire Hospital, Sheffield. Dr Nair indicated that he tended to abbreviate his name to Dr Siva Nair.

He indicated that HSP affects 1.3 in 10,000. He has been reviewing papers published about HSP, and identified that he had found 356 papers published since 1971, 142 in the last 10 years, and only 11 relating to treatment.

The databases he listed were:

AMED, http://www.ebscohost.com/corporate-research/amed

EMBASE, http://www.embase.com

PsycINFO, http://www.apa.org/pubs/databases/psycinfo/index.aspx

BNI, http://www.library.nhs.uk/help/resource/bni

CINAHL, http://www.ebscohost.com/academic/cinahl-plus-with-full-text

Dr Nair then went on to describe three treatments: FES, Botox and Intrathecal Baclofen, focussing on the latter.
Intrathecal Baclofen uses a pump and tube to deliver the muscle relaxing drug directly to the spine. One of the main issues is that it is very easy for the pump to deliver too much or too little drug. As an option, it is not one to be considered lightly.

Dr Nair posed the question why were there so few papers on treatment? A comparison with MS (I think) shows that about half of the papers are to do with treatments. One factor is that there are many groups who are actively influencing research there. He suggested that there was no reason why this couldn't be true for HSP as well. There were four strategies he suggested for getting involved;

1 - Participate in consultation events.

2 - Collaborate - get involved in research

3 - Control the research by selective funding

4 - Control the research by becoming a member of a steering committee.

Dr Nair concluded by describing a research programme which he is currently seeking funding for. He wants to look at the use of FES in the community. Most FES research has been done in the lab and Dr Nair wants to look how it works for users in normal day to day use. He plans to measure walking, falls, quality of life etc. at the beginning, then give either FES or an AFO for 12 weeks, re-measure, switch treatment for a further 12 weeks, measure again and understand what people prefer. The study will take place in Sheffield and Salisbury. He will contact the HSP support group when funding is secured. He will be seeking participants.

Promoting Walking Ability (Alison Clarke)

Alison Clarke is a physiotherapist from the Northern General Hospital in Sheffield. Alison began her presentation by giving the four things which a person needs to be able to walk:

1) Each leg must be able to support the whole weight of the body.

2) You must be able to balance on one leg

3) You must have sufficient muscle power to be able to swing the leg & trunk forwards

4) You must have the ability to swing the leg forwards.

Alison observed that for HSP the main problem is not being able to swing the leg forwards, there are problems bending the leg, with swinging it forward, and with heel strike. People with HSP bring their weight forward, with excessive trunk lean. As a result people with HSP have short steps, walking is slow, requires lots of effort, and various compensations are made. Several people end up falling over backwards.

HSP affects the gait because the condition affects the muscle tone. This can cause pain, bring on joint stiffness and reduce balance. When muscles are not used they become shorter and become weak.

When you have some walking issues, this is the time to have a review with a physiotherapist.

Alison then went on to discuss walking aids. These include:

Sticks/poles

Crutches/gutter crutches

Walking frame

Rollator/gutter rollator

Wheelchair

FES

'Gutter' refers to the type where you would put your forearm into a 'gutter' rather than hold just with the hand. Alison observed that with frames and rollators it is important to consider choices of seats, handles and ferrules.

Sticks and poles are used to aid balance rather than support weight. Nordic walking poles can be useful as they keep your body upright and increase your momentum. Having two of these aids is better than having one as two aids will keep you more upright. It is preferable to use your own muscles and balance to walk and move, but **considering using aids is not negative.**

It is important to choose your aids carefully, they should put less stress on your posture, give you less pain and give you more endurance. More importantly, perhaps, is that aids improve your aesthetics - people who use aids look more like an average walker than those who don't use aids.

Aids also give you more access to places, can help you at work or at leisure, and give you confidence. They can be a positive thing. If you move with your own muscles you will maintain the quality of your muscles and balance, and using aids will reduce this. Conversely you need to use aids when you cannot do what you want to, perhaps because you are falling or tripping. Therefore, its a balance - you will need to spend some time maintaining your muscles and balance, but also use appropriate aids when you are looking to enjoy yourself. Elbow crutches can be a good choice.

There was quite a bit of discussion around this balance between aids and self moving after the presentation. Generally people in the room felt they should have started to use aids earlier.

Many thanks to Adam Lawrence for summarising the above AGM presentations