



FSP Newslink

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

Issue 11 – May 2007

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

Chilling out on the Canals

Carolyn and I are just back from a week on the canals, and I thought I'd share with you how this works for us. We own a one twelfth share in a canal narrowboat. This allows us to have at least three weeks holiday during each year. In addition there are occasional opportunities for further time on board.

Obviously this is mainly a self-catering holiday, but you do have the opportunity to moor beside canal side pubs or restaurants and then stay overnight!

The holidays are very relaxing and we manage usually to have at least one other able bodied person on board to help with locks etc. Our boat is a six-berth boat, 58ft long and is called Somerton.

Having a shared boat makes the cost more affordable (running costs being split among the twelve owners) and we still get three holidays each year. There are several schemes in the UK which allow different forms of shared ownership - the one we chose is called OwnerShips [www.ownerships.co.uk]. In this scheme the company sets up the initial



arrangements, starts to sell shares and commissions the building of the boat.

Once the boat is launched it is the property of the joint owners and the company is retained (for a fee) to manage the running of the boat (maintenance, emergency callout, annual meeting of owners etc.). Approximate costs for a share in a new boat is £5K to £6K. Our share of the annual running costs during the six years have varied from £450 to about £1000 depending on what work the owners agree to have done. Additionally each week on the boat cost £70 -£90 for diesel, pump-out

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New Website:
www.hspgroup.org



of the loos (two on our boat) and a valet clean at the end of the week.

The boats are luxurious (shower/bath/TV/fridge/cooker/central heating/wood burner/microwave). Every owner looks after the boat when on board so it is kept in good condition. Newest boats also have washing machines and dishwashers!!

Carolyn has found it OK to get around inside the boat (no wide open spaces) and as three of the original owners had some form of mobility impairment some additional brass handles have been fitted in strategic places (bath, steps etc.). We take Carolyn's small dismantlable scooter for her to use when we can find a flat enough towpath and she uses it on most holidays to great effect.

Some boats are now being fitted out for people with disabilities - so it is worth checking these out if you are interested in future holidays on the canals! We'd recommend them!

Dave Harris

From the Chair

Hello Everyone

I open again on our weather. How strange; we haven't had rain for 5/6 weeks and the local parks are starting to look a little distressed. I drive out into the local Essex villages and the fields, trees, shrub lands and hedgerows are a flourishing green. With this extreme heat so early, does it mean a seriously hot summer?

So to some really good news, Mike Fawcett is now out of hospital. When we last spoke he was planning his next world tour, and that has to be a good sign. He would most certainly like to come to conference and can get himself to Leeds area if anyone could help him on from there.

I must say just a few words about the Jane Bennett's meetings at Milford in Surrey. I do get so much positive feed back and eagerly await for the next one. This brings me to our AGM and conference (agenda shown on page 8). We have a super day organised, time to tell you about the committee's activities, three speakers, a super buffet lunch

and of course the key to it all, ourselves. If you are new, it is a must. If you haven't yet attended, it is a must. If transport is a problem please let us know and we will see how we can help out. Ian tells me some 45 plus of us have already booked in, thank you, let's make this our biggest yet.

Wait for it. What a great success the Service Call system [www.service-call.net] to access assistance at petrol stations has been. I have used it mainly at Tesco's outlets where they now know me. On one single occasion I tried it out in an Essex village; no problems. It may be worth considering using it during a working day time period. In an evening when there may be fewer personnel this service invariable causes one service point to be closed.

Lastly, annual subscription forms will be coming round shortly. Could I ask you to deal with this promptly? It would be so nice if we could spend less time on chasing and more time on projects to benefit our members.

I look forward to meeting you all in Warwick on 16th. Have a safe journey.

David Pearce

From the FSP Helpline

Hi Everyone,

Just a few lines from me, I hope you are all enjoying this beautiful weather, I think we all feel much better when we see the sunshine. Please feel free to give me a call if you need to, it is really good to know how you all are, and how our chats develop. I look forward to seeing you all at AGM, especially as it is so good to put a name to a face.

Do please try and attend if at all possible you will all find it beneficial, and will meet many friends new and old.

Stephanie xxx

The Secretary's Spot

The timing of this issue of Newslink did not really allow us to give you enough formal notice of the AGM so this year we decided to send a separate letter. I look forward to seeing many of you again at the meeting which is now only about a month away.

Some members have started asking about renewal forms. We have this in hand and the forms will be with you before too long - so don't worry.

You may remember we had some problems earlier in the year with our web site. This led to some of the most recent information being "lost". The areas most affected were those containing the on-line copies of NewsLink and the messages in the users Forum area. The bad news is that there is not a sensible way to recover the user Forum data. The good news is that to prevent this happening again **we have now created a new site at <http://www.hspgroup.org>** If you attempt to visit our old site you will notice that you are now automatically redirected to the new site.

If we run a forum on the new site then those who wish to use it will have to register again on the new site - again we'll let you know when the time is right. *Dave Harris*

Useful Information

Naidex 24 –26 April

NEC Birmingham

The ultimate event for homecare, disability and rehabilitation.

Armed with my fast track entry pass I set myself off to see just what Naidex had to offer. Many years ago I attended the NEC as a representative for my company and had forgotten just how vast the complex is. We were however well sign posted and after some three hours (one stop included) I found myself in the undercover parking immediately adjacent the main entrance.

I subscribe to a number of mobility focused editorials and so a lot of the companies were already known to me. Most noticeable the array of exhibitors certainly covered every aspect of their running theme, (as above). So I set myself a plan to avoid missing

anything of value. Quickly I came across my scooter company and ordered solid tyres (numerous flat tyres in Epping Forest I can do without). Shortly after, Coloplast who had presented incontinence aids at Jane Bennett's Milford event the previous Saturday, came into view. I had some additional questions for them.

Some months ago Debbie Best introduced me to The Vibrogym, a vibrating platform which targets specific muscle groups to build, strengthen and **increase flexibility**. I have been using one in my local Ladies Hair Dressers but need more time to evaluate any change. I'll update you all on that later. However, a competitor, namely Power Plate also had a stand. I have details of their local operator and will pop in for comparison purposes.

Vibrogym: www.bettervibrations.com

Power Plate: www.powerplate.co.uk

As one gets into conversation it became obvious that the special Family House exhibit was going for the "most disappointing stand" of the show award. My condition is moving on and I felt this would be of particular benefit. However adaptations seemed little more than basic common sense with very little appearing to be new.

Below are details of products I came across in the afternoon and feel they might benefit some of us: A complete list of exhibitors is available if you have a particular interest.

Heat in a click: Pads enabling heat to be applied specifically to cold areas of the body. www.heatinaclick.com

Musmate: A series of elasticated straps and belts helping to recreate the mechanics of walking with particular attention to foot drop. I have ordered one and can't wait to **get it on**.

www.musmate.co.uk

The Neptune bath lift by Mountway: Probable the least obtrusive of bath lifts ever. www.mountway.com

PDQ International: Makers of the Powertrike which converts a wheelchair into a seriously fast electric tricycle.

www.pdq.tv

David Pearce

Urinary Urgency, Etc.

I think that nearly everyone in our membership appreciates that the spasticity of our lower limbs affects also, to a varying degree, our bladders, bowels and other nearby parts of our toiletry output (if you'll pardon the phrase).....and that that dysfunction will increase slowly with age.

Because of my own utilisation of the urinary drainage system (*and the immense relief and benefit that I obtain from it*) I wanted a representative of the company Coloplast, who produce the relevant items, to address a presentation to one of our meetings.

I, and my colleagues of Region 1, were very pleased with the talk organised by two of their nurse-representatives on 21 April at Milford, Surrey, one of whom (Claire Finch) has produced the following article. I am delighted to see that she has provided a contact number, so that any of you may telephone her and discuss the facilities and products and their availability via prescription, according to your needs and wishes.

John Moore

Coloplast is delighted to be celebrating its 50th Jubilee year in 2007. The Coloplast company goal is to help make life easier for our customers and the company is proud to be a premier supplier of continence products and services across the UK. From humble beginnings all those years back in Denmark, the company has always had the end-customer as its first priority, and this philosophy continues today.

On 21st April we were pleased to attend the Hereditary Spastic Paraplegic Society Group forum in Milford, Surrey. Daniel Waterman and Claire Finch represented Coloplast, with an aim to demonstrate the wide range of continence products available, and relate these first-hand to the everyday needs of members.

Coloplast has recognised through its supply of Ostomy products, the value to patients of Home Delivery, known to our customers as Charter Healthcare. This service is also

available to our Continence patients, ensuring an efficient, discreet and convenient way to get products directly to your home within 24-48 hours. The service is free, and with the support of the Charter Healthcare Customer Care team, help is at hand 6 days a week.

Daniel summarised the revolutionary Conveen Optima sheath, demonstrating its application and attachment to a leg bag collection system. This product has won an award for innovation, with its discreet packaging and unique features. Conveen Optima has been designed to provide the user with the opportunity to carry on with everyday activities whilst maintaining dignity and confidence. The product can also prove to be an effective alternative to wearing absorbent pads, which can be often be seen as undignified and uncomfortable.

For those who use a wheelchair, Coloplast has also devised a specialist range of leg bags, the knee bag which has a capacity of 1.5 litres, and the Syphon bag, which is strapped to the upper thigh. Both products can promote independence that may not be possible through the use of a standard urine drainage system.

As an alternative to an indwelling catheter, intermittent catheterisation can, on occasions, be an option. Our range of SpeediCath Catheters cater for all needs, including the SpeediCath Compact which has been designed specifically for ladies. It is important however to have a full assessment from a healthcare professional to check suitability prior to changing a product.

Finally, thank you for inviting Coloplast to attend your meeting, and should any member wish to discuss our product range or home delivery service (Charter Healthcare) in more detail, please feel free to contact Claire Finch on 07810 057 675.

Written by Claire Finch

Sales Specialist
Urology and Continence Care Division
Coloplast UK

Daily pill to beat genetic diseases

A pill that can correct a wide range of faulty genes which cause crippling illnesses should be available within three years, promising a revolution in the treatment of thousands of conditions.

The drug, known as PTC124, has already had encouraging results in patients with Duchenne muscular dystrophy and cystic fibrosis. The final phase of clinical trials is to begin this year, and it could be licensed as early as 2009.

As well as offering hope of a first effective treatment for two conditions that are at present incurable, the drug has excited scientists because research suggests it should also work against more than 1,800 other genetic illnesses.

PTC124 targets a particular type of mutation that can cause very different symptoms according to the gene that is disrupted. This makes it potentially useful against a range of inherited disorders.

The same drug could be given to patients with Duchenne muscular dystrophy, the most serious form of the muscle-wasting condition, cystic fibrosis, which mainly affects the lungs, and haemophilia, in which the blood does not clot. It can be taken orally, and safety trials have not revealed any major side effects.

“There are literally thousands of genetic diseases that could benefit from this approach,” Lee Sweeney, of the University of Pennsylvania, who is leading the research, said. “What’s unique about this drug is it doesn’t just target one mutation that causes disease, but a whole class of mutations.”

In most genetic conditions, between 5-15 per cent of cases are caused by a defect called a “nonsense mutation”. Genes are instruction manuals for cells to make proteins, but nonsense mutations in effect introduce a command halfway through that stops production. The kind of protein disrupted determines the nature of the disease.

In Duchenne muscular dystrophy, for example, the protein necessary for normal muscle development is not made, and the fatal wasting disease is the result. In haemophilia, it is the gene for the clotting agents factor VIII or factor IX that is disrupted.

PTC124 works by binding to a part of the cell called the ribosome, which translates genetic code into protein, and allows it to ignore nonsense mutations. The gene can be read straight through and a normal protein is produced.

The beauty of the drug is that it should be useful with any disease caused by a nonsense mutation, no matter what its outward effects. The error is not corrected, but ignored. Patients would have to take the pill throughout their lives.

PTC124, which is made by PTC Therapeutics, has been staggeringly successful in animal models. A study published today in *Nature* shows that in mice with a nonsense mutation that causes Duchenne muscular dystrophy, the drug starts dystrophin production and restores their muscles to health.

The drug has passed safety trials in humans, and the results of phase-two trials on cystic fibrosis and Duchenne muscular dystrophy will be published shortly.

About 13 per cent of patients with Duchenne muscular dystrophy have a nonsense mutation and should respond to the drug. It would not be suitable for treating different mutations in the dystrophin gene, or diseases not caused by nonsense mutations.

Other diseases that can be caused by nonsense mutations include beta thalassaemia, a blood disorder, and Hurler syndrome, in which children’s mental and physical development stops and most patients die by the age of 10.

Mark Henderson, Science Editor The Times

© The Times, London April 23rd, 2007

Members' letters

Unfortunately, we have not received any letters for publication in this newsletter. I believe that to a certain extent, the FSP Newslink can only be as interesting as the content we receive from our members. Please write in and tell us anything that you feel may be of interest to fellow members.

Material for publication in the Newslink should be sent to:

Ian Bennett
37 Wimborne Road West
Wimborne
Dorset
BH21 2DQ

I look forward to some interesting reading!

Regional News

Afternoon Teas Region 1 & 2

Our twice yearly Afternoon Teas are as much about catching up with friends as they are for sharing information and gaining help. Members, their family and friends, know that there will be someone in a similar situation to talk to and empathise with and if not knows someone who can. We have also benefited by being 'adopted' by members not living in either Region 1 or 2. Ian Bennett, the Membership Secretary and Region 4 Co-ordinator rarely misses joining us. We also welcome Stephanie (Helpline) and John as well as John and Stella, both from Region 3, who have adopted us and regularly attended.

I have often been asked why we have our meeting in Milford Surrey. The facilities at the Day Centre work well as it is disabled friendly. The location is almost central for Regions 1 and 2, plus it is not too far from the A3. For those that cannot attend due to lack of transport there are members who regularly offer lifts. There are a number of members who already car share/give lifts to those from their locality. A journey is often more pleasant if you have someone to talk to.

The format for the afternoon follows a similar pattern.

The first hour is given for Carers and family members to have time to discuss life with

those of us who are affected by HSP. Michael tells me that this works well as an information sharing exercise (and a chance to moan!).

The second hour is given either to Speakers or an Open Forum, sometimes a combination of both. Speakers have varied from companies, such as Coloplast to a chair exercise group such as Extend. Following their attendance at our last meeting I understand Clare, from Coloplast, has written a piece for the newsletter. John recommended and organised Coloplast's attendance. We always welcome suggestions from Members for speakers or discussion topics.

Dr. Jon Marsden, a regular speaker at our AGM, has joined us at a previous meeting to coerce 'guinea pigs' to help in his study. Some of our members, including Michael and I, took part in his study at the National Hospital. This worked well as I could attend as an affected person and Michael as a control; real teamwork!

We are sorry when members are unable to join us, but there is the lovely dimension that we still have contact with them. When we receive messages, letters and e-mails from members there is usually an update on their well-being and family news.

The continued contact with family members of deceased members is much appreciated and the group is enriched by their contributions. The group is for those affected by FSP/HSP, but for many our partners and family are a vital part of our lives and are equal members.

Thinking of the future. Those that do not join the Carer's group use this first hour to catch up and reacquaint. As many of us are finding out which gene is causing our FSP/HSP condition the suggestion has been made that we use this time to discuss our similarities and differences. We all have many similarities, but following open discussions, especially after Dr Marsden's visit; there are also a lot of differences. This could be the opportunity for those with the same affecting gene to get together and discuss symptoms.

The last hour is given to refreshments, raffle and farewells. We must say a big thank you to my parents, Ronald and Katie, who always

provide the delicious Afternoon Tea. The timing of the Afternoon's was devised to ensure there was time for Saturday morning tasks to be completed before making the journey, and not being too late finishing, especially with the dark evenings of November. This last meeting was typical of others in that we were 'pushing' people out the door at the end, only to find they stayed in the Car Park chatting for about another hour!!

With warm wishes, *Jane Bennett*

H.S.P. Group – Region 9

The Region nine group covers most of Northwest England from Shropshire to the Scottish Border and West of the Pennines.

Since joining the national group and meeting some of you in Devon last autumn, I have been trying to arrange a meet in this region.

There are some difficulties arising from the size and geography of the region. It is not easy to choose a place convenient to all, and to start with, it might be best to arrange a meet somewhere along the M56 axis between Manchester and Chester, because it will be easier for those in Manchester and Merseyside to come. The problem is where?

It might be then good to have a meet in Preston because that has a lot of motorway, train and coach links with a wide area. Again the question is where?

We need somewhere that is easy access to get to and get around inside, available Saturday or Sunday afternoon – and affordable. I don't expect anyone to stay overnight.

The problem with HSP (amongst others!!) is that not enough of us see the same consultant, and the condition is not widely known – or appreciated. We know that there are lots of forms of the condition, and that two strains have been identified at the National Hospital in Queens Square. We know that some people have the condition for no traceable reason, and some people are identified with HSP because there is no other 'slot'. We know that some people are initially diagnosed with MS or that relatives are diagnosed with MS and later re-classified as

HSP. This is a major issue as HSP is not a life limiting condition.

I would like to hold a meet-up with some folks to look at why we are where we are. Initially I want to do this on an informal basis, and maybe later involve some professionals.

I would appreciate ideas and suggestions, and anyone is free to call me. My email address for HSP stuff is mikeregion9@fsmail.net

Once we have chosen the venues, maybe there might be some 'cross boundary' interest as the M56 area is easily accessible from Wales and Shropshire, and Preston is easily accessible from Yorkshire. So maybe we can 'pool' the expertise in the three great medical schools of Leeds, Manchester and Liverpool.

Mike Cain

Flat 12 Highfield House, Beechfield Road,
STOCKPORT, SK3 8SF

0161 456 7531

Region 10

After a few years of hard work and enthusiasm, Ray Exley has decided to stand down as Region 10 co-ordinator. Thank you Ray for all the time and effort you have put in during this time. I have seen for myself how your very hard work has created strong friendships and bonds between members within your area. Well done!

If anyone is interested in taking on this position as co-ordinator for Region 10, please contact any committee member for further details. *Ian Bennett*

Notes from the South West

Seventeen members joined the May 12 Region 4 meeting in Ashburton. Dave and Tina came up from Redruth (our furthest SW members). We also had members from Bristol, Trowbridge, Weymouth and Wimborne - so the tradition of long distance travel was maintained!

Three of the attendees were new to the FSP Group - so there was lots of chat and information available to them. On this occasion we did not have a speaker, rather using the time to catch up on each other's news. After Ian started the meeting and we

all gave round-the-table introductions, the noise level slowly increased as conversation took hold.

Coffee/ tea and biscuits were served to table so that it would not get in the way of chat and all too soon the end of the afternoon loomed and people started departing on their journeys home.

Seven of the group were staying overnight and after a short interval we all met in the bar to commence the evening! A pleasant meal was enjoyed by all of us and we again repaired to the bar for further refreshment.

We now of course would need to travel home on the following day - this turned out to be the day on which the Ten Tors event (a few miles from where we were) was cancelled due to impending bad weather. The drive back to Weymouth on Sunday morning was very gloomy with severe spells of downpour. Despite this we all felt we'd had another successful event!!

Carolyn Harris

Forthcoming Events:

FSP Support Group AGM

Saturday 16th June

Trident Technology and Business Centre at Royal Leamington Spa (agenda below)

Mobility Roadshow

19th, 20th & 21st July 2007

Kemble Airfield near Cirencester.

Open 10 am daily Free admission Free parking www.mobilityroadshow.co.uk

New Members

We welcome the following new members:

Philip Makey Drifffield Region 10	Mr Steven Foot Sturminster Newton Region 4
Mrs Wilma Paterson Fife Region 11	Tania Williams Bristol Region 4
Karen Jordan Bristol Region 4	

Annual General Meeting & Conference 2007 Saturday 16th June

The Trident Technology and Business Park, Poseidon Way, Warwick, CV34 6SW

10.15 **Coffee & Welcome.**

11.00 **AGM**

11.30 **A short break**

11.40 **The Guide Post Trust: Christine Grimley**

"Who Cares for the Carers?" A light hearted look at the roles of friends and family as carers. Question and answer session.

12.25 **The National Hospital: Dr Val Stevenson**

Spasticity, What is it and what can be done for it: Spasticity can have a major impact on everyday activities such as walking, transferring, washing and dressing, and working? It is therefore essential to have an optional management plan incorporating effective use of treatment such as physical measures, medications, botulium toxin or intrathecal baclofen. This talk will discuss the importance of spasticity and what the individual and their health team can do to help. Question and answer session

1.25 **Lunch and the chance to mingle.**

2.40 **The National Hospital: Dr Jon Marsden**

Difficulties in Walking with HSP. Mechanisms & Rehabilitation. This talk will describe the results of a study that has investigated the cause of walking difficulties in people with HSP. Current and future studies that are looking at the effectiveness of interventions that address some of the difficulties highlighted in this study will also be discussed.

3.40 **Coffee and open Forum:** Simply relax, talk over the day or watch the DVD about Riding for the Disabled Association.

4.30 **Close**