PSP MATTERS

NEW PSPA STARTS HERE

WINTER 2019

BACK BY POPULAR DEMAND

5K FORPSPA

Will you be running, walking, pushing or skipping 5K to fight back against PSP & CBD this year?

MONDAY 18TH - SUNDAY 24TH MARCH

Join **#TeamPSPA** and run (virtually) alongside hundreds of others determined to raise funds to help people living with PSP & CBD.

In celebration of our 25th Anniversary, and as a special thank you for your support, we have made entry to this year's 5K for PSPA absolutely free!

Teams of 25 especially welcome!

Register for 5K for PSPA today at **www.pspassociation.org.uk/5KforPSPA**

WELCOME

As I reflect over the events of last year, I think one of my most extraordinary moments was when I looked over the attendance list for our first ever International Symposium on PSP & CBD, hosted jointly with Cure PSP, to see so many names of world-leading neurologists and researchers from 42 countries. The symposium was a strong reflection of the increase in research into PSP & CBD. On page 16 you can read about some of the research discussed at the event.

This edition of *PSP Matters* may look a little different to what you are familiar with. As you can read on our news pages (page 4) we have launched our new brand. Also, in this bumper edition, to mark the launch of our new Professional's Guide to CBD, we have put all CBD-specific information in its own section, *CBD Matters*. To go to *CBD Matters*, simply go to the back cover and turn the magazine upside down.

For some of us, the New Year is the perfect time for setting new challenges. If you would like to do something in 2019, why not take a look at some of our fundraising events on page 25.

Thank you for everything you do to support our vital work and I wish you a very happy and peaceful New Year.

Andrew Symons
Chief Executive

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PSPA NEWS

BOLD NEW IDENTITY AND WEBSITE

"Fab colours, so bold and strong. Looking forward to running in them!"
Supporter Charlotte Harmon

It involved a year of preparation and consultation, but finally in September we launched our new brand identity.

Branding can be an expensive business for charities, often costing many thousands of pounds. Last year we secured the free services of leading branding consultancy, Brandpie, through its charitable foundation. Brandpie spoke to a number of people affected by PSP & CBD, and, based on the feedback, created a dynamic and optimistic brand, made up of bold colours and a new strapline, "Starts here".

One of our greatest challenges is raising awareness, with around half of people misdiagnosed initially, and many health professionals unaware of PSP & CBD. To face these challenges, we need to build a strong brand that unites and brings together everyone affected by PSP & CBD. We hope our new brand will help us with our plans to bring more attention to PSP & CBD, secure better care and invest more in research.

The more people who join our community, the stronger we will be against the challenges we face. Thank you for your support. We can't wait to see many of you wearing our new colours this year!

OUR NEW WEBSITE

To coincide with the launch of our new identity we have created a website **www.pspassociation.org.uk** Designed to look as good on a phone as it does on a laptop, the new website clearly separates our key areas of work, allowing people affected by PSP & CBD to access information easily.

Our work on developing the website continues. It is really important we get any feedback you have on it and how it works. We will be collating all feedback and working with developers on improvements. Please email your comments to **info@pspassociation.org.uk**

ANNUAL REVIEW

Our review follows the journey of a single pound through the charity, so you can see where the money you raise goes. In 2017/18 thousands of you, our PSP & CBD community, turned your personal experience into action and raised over £1.188m for PSPA. Our annual report, details how the money you raised was spent in 2017/18 and what we did to support people affected by PSP & CBD. You can read our annual review on our website in the About Us section.

ANNUAL REPORT

THE JOURNEY SO FAR

Driven by his own devastating experience of watching his wife die from PSP, Michael Koe set up a charity from his kitchen table with the single goal of helping others affected by the disease.

24 years on PSPA provides vital information and support to people living with PSP & CBD and their families, alongside educating health and social care professionals so they can provide the very best care. PSPA also funds research to help find a cure.

Michael Koe's work continues thanks to the thousands of people who have also seen their lives changed forever following a diagnosis of PSP or CBD. Sadly, half of these people received the wrong diagnosis, before being eventually told they actually have PSP or CBD.

At least **5,000** people are living with PSP & CBD in the UK, but this number could be more than **10,000** as many are misdiagnosed with other conditions. Life expectancy is typically between five to seven years, but can be as long as 10 years from onset of the symptoms.

04

THE JOURNEY SO FAR

YOU ARE TURNING YOUR EXPERIENCE INTO ACTION
People with PSP & CBD across the UK, their families, friends and colleagues are turning their devastating personal experience into action.

Our community is made up of people with PSP & CBD, their families, friends, neighbours and colleagues, alongside health and social care professionals and researchers. We are a movement. We are united in the fight against PSP & CBD.

HERE IS THE STORY ABOUT WHAT YOUR ACTION ACHIEVED IN 2017, TOLD BY FOLLOWING A SINGLE JOURNEY OF A POUND.

05

PLEASE TELL US WHAT YOU THINK

There have been a number of big changes recently. We would really like to know what you think to our new look annual report, as well as our new brand. Please could you spare three minutes by visiting our website and completing the questions in the About Us/Annual Report section.

£70,000 RAISED FOR THE NEXT STAGE OF PROSPECT

Last autumn we asked if you would be willing to donate to the next stage of the biggest, most ambitious PSP & CBD research project to date. Thousands of you donated and we are thrilled to say our appeal has raised over £70,000. Thanks to you, our researchers will be able to expand recruitment to involve more people with PSP & CBD, analyse the huge amount of data gathered so far, and better understand disease progression and discover diagnostic markers.

PSPA TURNS 25 THIS YEAR

2019 is a very special year for PSPA. It was 25 years ago when our charity was first set up by Michael Koe. Since then it has grown to a charity supporting thousands of people living with PSP & CBD. This year, to mark this special anniversary, we plan to increase our activity by a factor of 25. We aim to support 25% more people affected by PSP & CBD. We plan to recruit 25 more education and support worker volunteers, and, we are aiming to target 25 new neurologists. To help us achieve this we need your help. If you would like to volunteer or are planning to mark our 25th birthday please email **info@pspassociation.org.uk.**

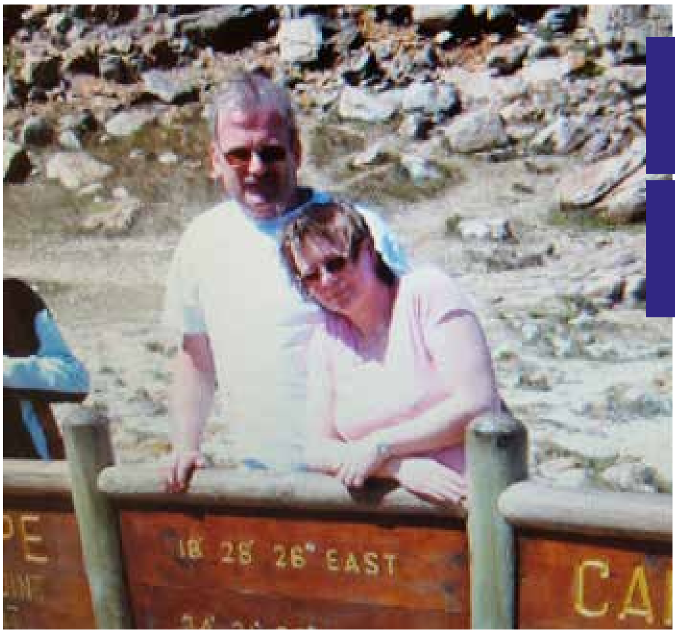
UNDERSTANDING DISEASE PROGRESSION

Our Sara Koe Clinical Research Fellow, Dr Edwin Jabbari, has published the results of his work in the journal *Annals of Neurology*. Dr Jabbari originally set out to improve understanding of why some people with PSP experience faster progression than others, and his new paper provides part of the answer. Ultimately, this area of work will help neurologists give families a more accurate prognosis, as well as making clinical trial results easier to interpret.

Dr Jabbari and colleagues at University College London looked at the genetic differences between patients with typical PSP compared to patients with slower progressing forms of PSP. In total they studied 600 PSP patients and found that variation in a gene called TRIM11 was an important determinant of the characteristics of an individual's PSP. Interestingly, this gene may play an important part in the nerve cells' waste disposal system, called the ubiquitin proteasome system (UPS), which is involved in the breakdown of misfolded proteins, including tau.

While follow up work needs to be done, including looking at the impact of this gene in other neurodegenerative diseases such as CBD, it is possible that the UPS could be a target for future drug development.

A significant portion of the 600 PSP patients came from our PROSPECT-UK study, which so many of you have participated in; the clinical data and DNA samples that you donated were central to the success of Dr Jabbari's work. Dr Jabbari said: "My study wouldn't have been possible without PSPA and all of its supporters. I'd like to say a massive thank you to you all!"



MEET ELIZABETH

"It is so important for health professionals to be educated because when they are, it really makes a difference when it matters the most."

Elizabeth Reade's husband is living with PSP. Elizabeth talks about the changes they have had to face as Robert's PSP progresses.

"Standing up to give the father of the bride speech at our daughter's wedding, it was clear something was wrong. Robert's speech was rambling, and a bit strange. Now of course we know why, but at the time we had no idea what was happening in his brain.

"Looking back the signs were there, but we just didn't realise how serious it was. My husband had always been very fit and active. He was a sociable man in his early 50s who had a high-profile job that took him around the world. Around six years ago his personality started to change. He didn't want to socialise. I noticed he started to drive the car too close to the kerb – something he had never done before. I thought perhaps he was depressed and under too much pressure at work. Then, he became reluctant to travel abroad. For a man who spent so much of his time travelling, this was very odd.

"In 2014 he started to experience balance problems. I also noticed how his foot would catch on the floor when he walked. By the end of the year he was falling over quite regularly. His personality had completely changed. He was certainly not the man I married. I frequently tried to get him to go to the doctor, but he always resisted. After one particularly bad fall I made him go. That is when he told me his work had made him go to occupational health for an assessment. They clearly also noticed something was wrong. He underwent a conventional dementia test that was inconculsive. Occupational health simply said something was wrong, but they couldn't identify what it was. They recommended he saw a doctor. Finally, in March 2015 we got him to see a GP. The GP was very good. He said he didn't know what was wrong, but that he needed to see a neurologist. Robert was sent for an MRI and nerve conduction tests. During this time, the troubling thing was Robert didn't seem worried at all. In May, we were told the devastating news Robert had PSP. He was 55.

"By the time of his diagnosis, Robert was no longer able to forward plan. It has been very difficult. All the way along I was hoping he had Parkinson's. How funny to hope your husband has Parkinson's disease.

"HOW FUNNY TO HOPE YOUR HUSBAND HAS PARKINSON'S DISEASE."

“WHEN YOU SAY YOUR PARTNER HAS CANCER PEOPLE UNDERSTAND HOW SERIOUS IT IS. WHEN YOU SAY YOUR HUSBAND HAS PSP THEY JUST LOOK AT YOU BLANKLY. YOU ALWAYS HAVE TO EXPLAIN IT.”

“Nine months after his diagnosis, I left my job as a radiology manager for North Tees hospital. I started to work part-time, three days a week. Robert was medically retired. Robert and I were planning to work until we were 60, but now due to the dent in our pension savings I will have to work much longer. People often don’t realise the financial implications of getting a disease like this when you’re still working. All your plans for the future are suddenly dashed.

“Robert gets PIP, but we had to appeal for him to get the full amount. PIP now funds most of his care bill. He has carers in for four visits a day, seven days a week. We now have CHC funding which funds half his care. I don’t think the assessors have a good understanding of PSP and what it means. The guidelines say they shouldn’t be disease specific but I think they are.

“When you say your partner has cancer people understand how serious it is. When you say your husband has PSP they just look at you blankly. You always have to explain it. Some days that’s okay, but other days I just don’t feel I have the energy to go into details.

“Robert has been badly affected cognitively by PSP. His personality is so different. That is the most difficult bit. As a person he has totally changed and that happened early on. He can’t swallow, walk, struggles with his vison and has to have a catheter. His catheter is a pain because he gets one infection after the other yet can’t tolerate antibiotics.

“I try and take Robert out, but it’s very tiring for him. He always wants to go to the PSPA support group. I honestly don’t know what he gets out of it, but it’s something he wants to do. Afterwards, he needs at least two days to recover. Robert will be 60 at the end of May. He has refused a PEG feeding tube. From what I can understand, he doesn’t want to go into hospital. He is scared if he goes in he won’t get to go home.

“Accessing the support group and PSPA has helped. Simply speaking to other people who understand the illness is helpful. PSPA sent me some really useful information about CHC funding and has helped me prepare for the meetings.

“In the last few months since Robert has deteriorated we have had huge support from the GP and the district nurses. The district nurses have gone out of their way to find out about PSP. This makes a huge impact and to have informed and active health care professionals really helps. I understand why it is so important for health professionals to be educated about PSP because when they are, it really makes a difference when it matters the most.”

KATIE WITH HER UNCLE MICHAEL AND HER MUM ANNE

"MY UNCLE SPENT HIS LIFE HELPING OTHERS, SO IT IS NATURAL TO RAISE MONEY IN HIS HONOUR."

Not having a bike didn't deter Katie Hodder from embarking on a 100-mile bike ride through London and Surrey for PSPA. Katie decided to take on the gruelling challenge in honour of Prof Michael Hay, her beloved uncle, who is living with PSP.

"My Uncle Michael received his diagnosis of PSP in 2016, but he first became ill a few years earlier. It was put down to other things, sadly preventing him from getting an accurate diagnosis and access to the most appropriate care.

"Since his diagnosis, the deterioration has been rapid. Now, this former publisher and distinguished globetrotting professor and fun-loving family man, requires 24/7 care with the loving support of family and an incredible team of carers. Michael now lives in a home where my aunt visits every day to help care for him. He is bed bound and can't talk, but he still has a good quality of life. When we take him for a day out to the park, or back to his family home he gives us a thumbs up.

"Michael was a very well-known Professor and Dean at the London Business School. He is a brilliant man who has achieved many things during his career.

KATIE AND HER BOYFRIEND CHRISTOPHE

Watching him deteriorate is incredibly sad. We all felt so helpless. After experiencing the challenges in getting a diagnosis, we wanted to try and improve the situation for other people. Being a nurse, I know what support is out there for other conditions like cancer and heart disease. And, in contrast, how little there is for PSP. We need to raise awareness of PSP, and improve the care people, and their families receive.

"My uncle spent his life helping others, so it felt natural to focus on raising money for PSPA, as a way to help others affected, in his honour. Once I started fundraising, I soon discovered that it has enabled others to feel like they are doing something to help, by making a donation. After I posted my VirginMoney sponsorship page I received so many messages from people to say that donating made them feel like they were helping in some way.

"My uncle and I always played sports together, so I knew whatever I did to raise money had to be sports-based. It also needed to be a substantial challenge to motivate people to put their hands in their pockets. When I saw RideLondon, I knew that was the perfect challenge for me. I didn't even own a bike, let alone feel able to take on the 100-mile bike ride.

"I started training in March. The first step was getting used to my new bike – it looked so thin! Gradually over time I built up strength and distance. As a nurse practitioner, finding time to train was challenging, but at the same time it felt good to spend so much time outdoors.

"The next step was creating an online sponsorship form with Virgin Money. My target was to raise £450. I reached out to all my uncle's old contacts at the London Business School who came out in force to donate, smashing my initial target. I watched in disbelief as my total rose every day to a final £12,400. It was so lovely reading the kind messages people had written about my uncle. I also did some local fundraising and organised a pub quiz in Clapham. The local businesses were so supportive, along with larger companies who donated great raffle prizes.

"The whole RideLondon experience was brilliant. Even waiting to start in the rain at 6am it felt so special. All the riders were sharing their own stories about the charities they were supporting and why. Going uphill, the other riders would cheer each other on. When I crossed the finish line I burst into tears. It felt so good knowing I had done all this for a charity where every penny goes back to the people who need support. High on adrenalin we started to cycle home, but it all became too much and we ended up on the train!

"There are so many more reasons to do something positive in the face of PSP, than not do it. If it wasn't for my uncle I probably wouldn't be a nurse. He has done so much for me, so doing this for him is my way of giving back. The fact that the money I have raised could help fund a researcher for three and a half months is the best way I can show my uncle what he means to me and how grateful I am to him."

"THE WHOLE RIDELONDON EXPERIENCE WAS BRILLIANT."

KATIE AT THE FINISH LINE

“PEOPLE WORRY ABOUT GOING ABROAD, BUT WITH PLANNING IT IS POSSIBLE.”

TASNEEM AND MUNIR AKHTAR IN HONG KONG

Making the most of time together can be especially challenging if your loved one has PSP or CBD. In January, many people’s thoughts turn to holidays, but for people living with PSP & CBD, going on holiday may seem impossible. With the right guidance and some planning holidays can be enjoyed as Nadia Akhtar found out after her dad received his diagnosis of PSP.

“Dad had every single symptom associated with PSP. One of the hardest things to cope with was how his personality changed. He was very active and sociable, but looking back, a few years before his diagnosis, he started to withdraw. It wasn’t long before he bore no resemblance to the person we knew. Dad became very apathetic. It was very clear something was wrong and the changes in Dad could not simply be put down to ‘slowing down’.

“Dad experienced a series of losses, each one eroding a little bit more of his independence. Losing the ability to drive was a big issue for Dad. And seeing that helplessness was really hard. The falls accelerated. It was alarming, scary and upsetting.

“We managed PSP as a family. Both my sisters and I lived at home and helped Mum as much as we could. We agreed to stay with Dad on alternative Saturdays so Mum could continue to work. Work gave Mum a lifeline and an opportunity to get out. However, she did the bulk of the caring. She had to tackle the difficult behaviours as he could sometimes be very aggressive.

“Through securing help, we were able to keep Dad at home, and I was pleased we were able to look after him until the very end. We made the most of our time together and focused on creating memories we still talk about now. I’m so pleased we continued to have holidays with him. Every year we went abroad. We visited Germany, Corfu and did a tour of China, including exploring eight cities and a river tour, and, just before he died, we visited Oman.

“Dad loved his holidays and had a real sense of wanderlust. People worry if they can still go abroad, but with some planning it is possible. We researched disabled facilities and packed packages of food, including powdered custard, noodles, cake bars and basically anything he was used to eating at home, including his thickeners. We planned each trip with Dad’s routine and capabilities in mind. In Oman, we booked a disabled room and contacted the hotel before we left. With the advance notice the hotel was extra helpful with Dad. We also planned trips with rest stops so he could nap and we avoided overnight flights. Wheelchair assistance at the airport is a brilliant service and worked really well.

"As hard as it gets it is really important to try and focus on how life is worth living. We cared for Dad as best we could and miss him every day since he died."

PLANNING A HOLIDAY

With planning, many holidays can be made accessible. Here are some tips to help.

- **TRAVEL SPECIALISTS** Contact accessible travel specialists. Tell them about your needs so they can recommend the most suitable holiday.
- **AIRPORT ASSISTANCE** This can be pre-booked and arranged free of charge. Someone can help you from your taxi at the airport, at the check-in desk, escorting you to the aircraft and helping you to board.
- **MEDICATION** If you need prescribed feeds and fluid supplements on the plane, they must have the original label and packaging given by a pharmacy, along with a doctor's note to ensure they can be taken through security and check with your airline before flying, to see if any security restrictions apply.
- **TRAVEL INSURANCE** Many insurers cover PSP & CBD. For more information contact our Helpline.
- **PLAN AHEAD** Research restaurants, bars and places you want to visit. You might want to check your holiday destination isn't hilly and has good accessibility.
- **WHEELCHAIRS** Carry your wheelchair instruction manual with you. Airlines want to know the weight and dimensions of mobility scooters and electric wheelchairs at the time of booking.
- **CHARGING** An 'airsafe' plug can be purchased to inhibit powered wheelchairs on flights. This device plugs into the charging socket to immobilise the wheelchair. This can prevent damage, as security regulations may result in wires being cut or removal of the battery. Search for airsafe plugs online.
- **CARE** Some holiday destinations offer care services. The Disability Aid Trust help towards the costs of care workers.
- **INFORMATION** Medical staff may be unfamiliar with PSP or CBD. Take information about PSP & CBD with you, like our handy business card sized fact cards.

USEFUL CONTACTS

- **Accessatlast** 01772 814555, www.accessatlast.com
- **Accessible Property Register** www.accessible-property.org.uk
- **Limitless Travel** 0800 069 8060, www.limitlesstravel.org
- **Disability Aid Trust** 0800 028 0647, www.disabilityaidtrust.org.uk

For more information on taking a holiday please contact our Helpline on **helpline@pspassociation.org.uk** or on **0300 0110 122**

TASNEEM, NATASHA, NADIA AND MUNIR AKHTAR IN CHINA

"WE PLANNED EACH TRIP WITH DAD'S ROUTINE AND CAPABILITIES IN MIND."

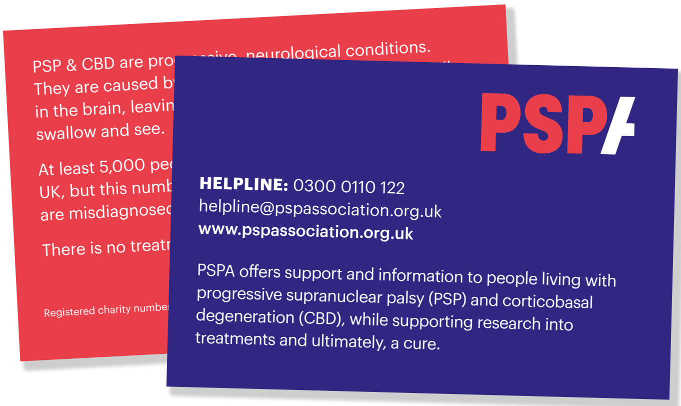
COGNITION AND COMMUNICATION

Cognition refers to our thinking or our mental processes of perception, memory, judgement and reasoning. For many people caring for a loved one with PSP & CBD, changes in their cognition and speech can be one of the hardest things. We asked Chloe Cripps, a Speech and Language Therapist specialising in supporting people with cognitive changes, to share her advice.

"Changes to cognition can impact on how quickly you are able to process what is said to you and formulate a response. It can affect your memory causing you to lose your train of thought in conversation. It can cause changes to attention, making it difficult to concentrate and follow a conversation.

"The impact of speech changes in PSP or CBD on communication is generally well recognised. However, the impact of cognitive changes on communication related to PSP & CBD is sometimes overlooked. How people's cognition is affected by their condition will vary hugely. Whether these changes are big or small, they often affect how a person communicates and how they interact with others.

If you are experiencing cognitive changes as a result of PSP or CBD there are things you can do to try to lessen the impact on your communication:



PSP & CBD are pro neurological conditions.
They are caused b
in the brain, leavin
swallow and see.

At least 5,000 peo
UK, but this numb
are misdiagnosed

There is no treatr

Registered charity number

HELPLINE: 0300 0110 122
helpline@pspassociation.org.uk
www.pspassociation.org.uk

PSPA offers support and information to people living with progressive supranuclear palsy (PSP) and corticobasal degeneration (CBD), while supporting research into treatments and ultimately, a cure.

- **LET PEOPLE KNOW** Let people know what's difficult for you. Some people carry a card which explains their condition and asks people to give them more time. If you find it difficult to do this yourself, is there someone who can let people know on your behalf?
- **TAKE YOUR TIME** If you take your time when you're trying to get your message across, you will be able to express yourself more easily.

- **JUST ONE THING** Do one thing at a time. Finish making the cup of tea before you have a conversation, pause the television if you want to chat.
- **MANAGE YOUR TIREDNESS** Being tired will make your cognitive symptoms worse. If you've not slept well, you may find conversations are harder. Communication may be harder at the end of the day when you are at your most tired. Try to arrange your day to ensure you plan social events when you are at your best.
- **LIMIT STRESS** If you are feeling worried or frustrated it will make it harder to put your thoughts into words. It may be better to start the conversation again later. Or perhaps ask 'can we start from the beginning?' Giving you time to take a breath and collect your thoughts?
- **WRITE THINGS DOWN** It may help to carry a notebook with key information in it. You may wish to ask people to write down what they have said to you.

WHAT CARERS AND LOVED ONES CAN DO TO HELP

Often it is what other people do that is the key to a person with communication difficulties being able to continue to be successfully involved in conversations. There are many things you can do as a carer, family member or friend to help.

- **FIND OUT** Ask them what's difficult so you can better understand what they are experiencing. Next find out what they find helpful. For example, do they want you to give them more time if they get stuck or do they like you to fill in the gaps?
- **CREATE THE RIGHT ENVIRONMENT** Try to reduce background noise, get rid of distractions and make sure you are face-to-face and have good eye contact whenever possible.
- **STAY RELAXED** If you can't catch what they're saying, try not to get frustrated. If you see them start to get frustrated try to adapt the situation to reduce this. You could go somewhere quieter, perhaps sit down face-to-face and give them your full attention.
- **TIME** Give the person time to respond. Take time to explain things and make sure it doesn't feel rushed.
- **BREAK IT DOWN** Breaking down what you say can be helpful. Rather than 'Paul and Jean are coming with the kids, then I think we'll go to that restaurant, the one around the corner from Bill.' Say 'Paul and Jean are coming. They're bringing the kids. We'll go to a restaurant. The restaurant around the corner from Bill's house.'
- **QUESTIONS** Questions can put people on the spot and are often difficult. Reduce the number of questions you ask. Adapt the questions you do ask, for example, give options or ask yes/no questions.
- **DON'T CONTRADICT** If there's confusion or someone misremembers, consider if it's necessary to correct the person or can you focus on the meaning behind what they are telling you, rather than the factual accuracy?
- **COMMUNICATION AIDS** If a person uses a communication aid such as a book with words, pen and paper or a text-to-speech device, make sure they are always available.
- **INVOLVE THE PERSON** Speak to THEM – don't think they're not listening if their eyes are closed. Don't think they don't want to answer if they can't answer immediately.

It is important to remember that everyone is different. We all have our unique communication styles and personalities. PSP & CBD affect people in different ways. What works well for one person may not work well for another. Ask for help. Speech and Language Therapists are experts in communication and can consider strategies that are most helpful for you.

CHLOE CRIPPS, SPEECH AND LANGUAGE THERAPIST

"I WAS SO NERVOUS ABOUT COMING BECAUSE OF FACING UP TO A DIFFICULT FUTURE, BUT I AM SO GLAD I DID. THANK YOU."

Over 80 people came along to our Family and Friends Day in Old Windsor. The day opened with a talk from Dr Ruth Lamb (see page 28) followed by an opportunity to ask our panel of health professionals questions on a range of subjects including diet, communication aids, benefits and how to access services.

The afternoon session included an excellent musical interlude from Sound Alliance and an update on the work of PSPA from CEO, Andrew Symons. The English National Ballet brought our day to a close with an excellent dance and movement workshop where people with PSP, CBD and their family members were able to join together and express themselves through movement and sound.

Family and Friends Days provide an informal opportunity for people living with PSP, CBD and their families to come together to learn more about the condition, recieve updates on research and the work of the charity.

COMING SOON

Please visit our website for details of our 2019 Family and Friends Days.

300 HEALTH PROFESSIONALS ATTEND OUR EUROPEAN STUDY DAY

Around 300 health and social care professionals joined us at our Study Day in October at the Royal College of Physicians, London. The event was sponsored by CBD Solutions Sweden, who presented the latest research developments in CBD.

Neurologists and researchers from across Europe presented the very latest updates on symptom management, research, end of life issues and the importance of multidisciplinary team working.

Alexa Wearden, Specialist Occupational Therapist, Hammersmith and Fulham Community Neuro-Rehabilitation Team was one of the professionals who attended the education event. Alexa said: "I work as part of a community rehabilitation team in Central London.

"The study day included updates on cutting edge research, recent developments in understanding the disease process, and sessions discussing how people with PSP & CBD may present in real-life. Having the opportunity to listen to clinical experts, who are also working in practice, was invaluable. One clear theme that emerged was the importance of community-based teams, such as our own, that can provide a holistic co-ordinated multidisciplinary approach.

"This study day provided an excellent opportunity to develop my own knowledge of PSP & CBD. The day finished with a moving film of a person living with CBD and her own journey as the disease progressed. It reminded us all how life changing this diagnosis is, and the need for us to come together as a community of professionals to ensure we are providing the best possible care. I was so inspired by the study day that I have already shared some of my reflections and learnings at our weekly multidisciplinary team meeting.

"Finally, my colleagues and I were particularly impressed by the range of resources offered by PSPA, and are excited about the latest guide on CBD and online resource (see page 30). I found this study day stimulating, challenging, inspiring and I am already looking forward to attending next year's event."

You can access the presentations from our website **www.pspassociation.org.uk/information-and-support/for-professionals/study-days/**

COMING SOON

We are working on a programme of events for health and social care professionals for 2019 including three mini Study Days in Wales, Scotland and Northern Ireland and our annual Study Day taking place in one of the major cities in England.

INTERNATIONAL SYMPOSIUM ON PSP & CBD SIGNALS A NEW ERA OF HOPE

Over two days, 220 leading scientists and researchers travelled from 42 countries to London for our first International Symposium on PSP & CBD, jointly hosted with CurePSP.

The event covered the latest research, from the basic biology of tau to immunotherapy trials, from PSP & CBD brain banks to patient registries, from imaging and fluid markers to genetics. Clinicians and researchers attending the event were united by a common aim; to better understand PSP & CBD and to provide better clinical care to people living with both conditions.

Young researchers who will be tomorrow's leading research figures were also in attendance, with 40 early career investigators presenting their work at the poster presentation sessions. "It is extraordinary to have both the leaders in research over the last 20 years, but also, dare I say it, the young generation of doctors and scientists" said Prof James Rowe, University of Cambridge. James continued: "What is evident from this gathering is the positivity and progress; better diagnosis and a real sense of optimism about future treatments."

Opening the event was Dr John Steele, who along with Dr Richardson first described and recognised PSP as a condition 55 years ago. John told the room full of PSP & CBD researchers: "I believe we can now find the cure. That is my optimism and your challenge. Solve this mystery of the last 50 years."

This first session focused on what researchers have discovered by looking at brains donated by people with PSP & CBD, with presentations by Dennis Dickson from the Mayo Clinic Brain Bank and Janice Holton from the Queen Square Brain Bank. The researchers paid tribute and thanked the patients who have donated their brains. They have provided an invaluable resource to researchers and contributed to a huge number of research papers globally, building on our knowledge of the role of tau and how it distributes itself in the brain.

Queens Square Brain Bank collects brains from all over the UK. Around 25 brains from people with PSP are donated each year and a smaller number of brains from people with CBD. Prof Holton outlined how many of these patients have been seen in the clinic at Queen Square, so researchers have a good amount of clinical data to match to the brain samples, making them especially valuable. This is particularly true for the brains of PROSPECT study participants, where vast amounts of information have been gathered. Any researcher in PSP & CBD can request tissue from the brain bank, with requests coming from all over the world.

"Just a few years back we'd have been lucky to get 15 people at a PSP meeting," said John Hardy, Professor of Neuroscience at University College London. "There is now much more clinical interest and drug company interest in PSP & CBD. People are building up to clinical trials as we get to know more about the diseases. I am a geneticist and this event has illustrated the amount of progress in genetics over the last ten years.

"We have known the tau protein is important for a long time. Now we can also see some of the other proteins we are finding are involved in degrading tau. This is interesting as it is starting to give us mechanistic clues which we might be able to use down the line, to develop new therapies. This is an era of hope. More people, more investment, more knowledge, more drug targets. It is a good time for PSP & CBD research and I hope over the next five to ten years we start to see new therapies come out of this."

Prof Adam Boxer, from the University of California talked about really interesting ways of increasing efficiency of clinical trials. Adam suggested the research community needs to learn from cancer medicine in order to move more quickly towards a cure.

At the heart of this will be more clinical trials. Adam described innovative ways to increase the efficiency of small trials in rare diseases, like PSP & CBD. It was also interesting to hear about a trial of a sleep medication where participants never even had to visit the clinic to take part. Adam outlined how we need to get to a stage where all people living with PSP & CBD are able to take part in a trial if they wish, even if housebound.

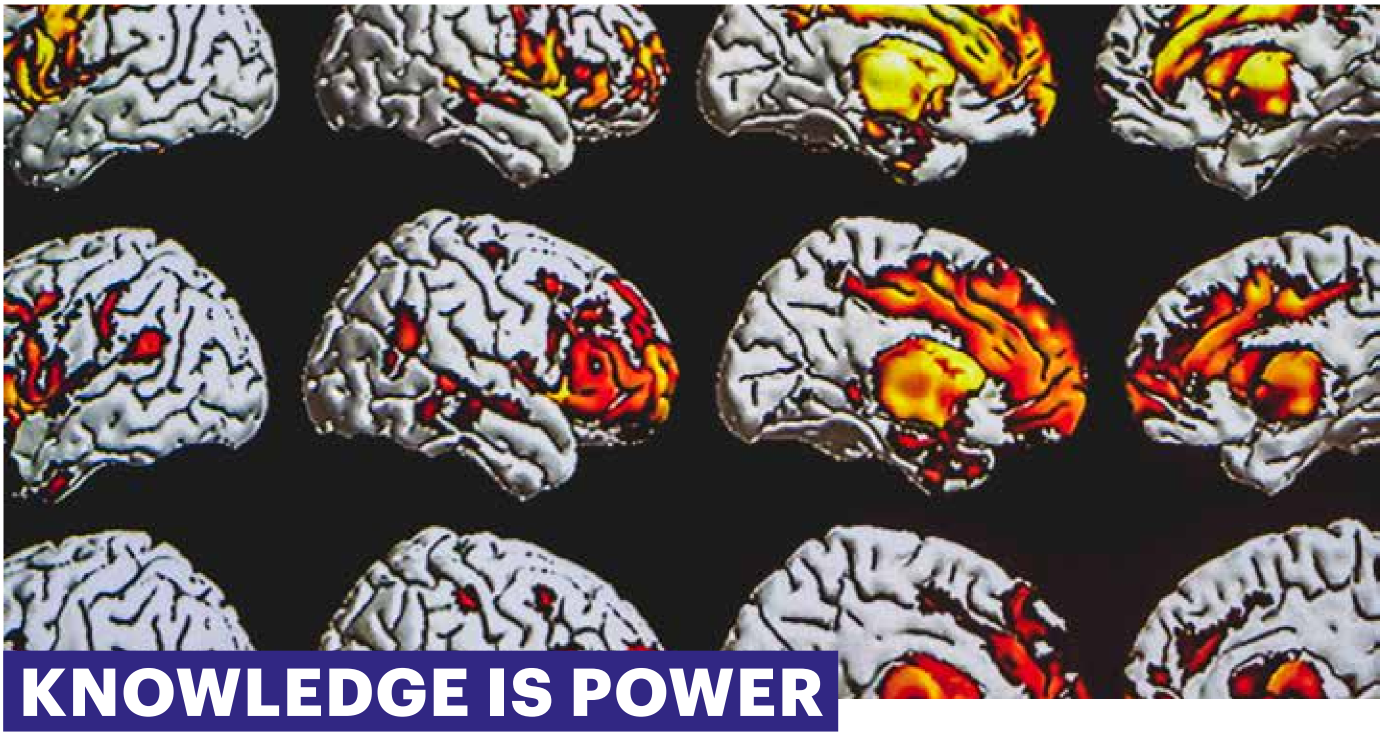
"IT IS A GOOD TIME FOR PSP & CBD RESEARCH AND I HOPE OVER THE NEXT FIVE TO TEN YEARS WE START TO SEE NEW THERAPIES COME OUT OF THIS."

JOHN STEELE

Ultimately, effective treatments for PSP & CBD can only be developed through collaboration and the sharing of information. Many researchers spoke about the importance of events like this for making that happen. Shauna Yuan, from the University of San Diego, said: "The symposium fostered a collaborative environment bringing together the research community. I felt very privileged to be part of this well-run event.

"The talks provided a perspective of how much the field has grown and provide much hope for what's to come. I found the research talks to be top-notch and cutting edge. I was able to make connections with other researchers that will have a direct impact on my work going forward."

For the young researchers, the event helped them build up useful connections. Dr Wael Ibrahim, Egypt said: "I am a young doctor from Egypt specializing in movement disorders. I chose this event to present my work in order to get in contact with eminent doctors and colleagues working in PSP & CBD to benefit from their ideas and research."



KNOWLEDGE IS POWER

Science writer Maria Tennant shares what she learnt from the event. Maria asked to attend following her mum's diagnosis of CBD. So, for Maria, the research presented felt very personal.

"My mum was diagnosed with CBD earlier this year. As the condition is so rare, we don't know that much about what we are dealing with. I want to understand more about it, so to have the chance to learn about both PSP & CBD from the world's experts seemed too good an opportunity to miss.

"I've been to many scientific conferences in my time, but this was the first time I had been to one that felt so personal. I wasn't quite sure how I would feel about listening to the talks – and having my mum and the implications for her in the back of my mind. But hearing about the exciting research going on around the world has made me feel like I am doing something to help. It has got me thinking about questions we can ask the doctors, and about things Mum should be involved in.

REVEALING WHAT'S HAPPENING INSIDE BRAIN CELLS

"The morning session was all about the basic science of PSP & CBD - about what is happening in the brain, right down to the individual cell level. Patients who kindly donate their brain to research after they die are having a huge impact on what we know about these conditions. It is revealing much about the genetics and pathology of both PSP & CBD, and how different regions of the brain are affected. When a patient donates their brain, it is stored in a 'brain bank', and researchers go through a strict approval process to access brain tissue samples for their research.

"Both PSP & CBD fall into the same category of brain conditions – the tauopathies – where the protein tau becomes faulty and builds up within the brain. Researchers have found this tau build-up affects cells in the brain differently, and the areas reflect how a person is affected physically and mentally. For example, cells in the front of the brain are affected in CBD, which includes the areas controlling movement, whilst cells elsewhere are affected in PSP.

"Helen Ling from University College London (UCL) explained how it's vital we work out the earliest changes in the brain that lead to PSP & CBD before any symptoms develop – it might reveal new ways to intervene earlier. Her work is discovering that tau levels in early stage disease are much lower than late stage. Her team has worked out the path that most disease cases follow through the brain, but more aggressive diseases move along it quicker. But her work is revealing early brain changes don't always predict how the disease will progress, and that a process called inflammation could be involved.

GENES ARE IMPORTANT

“We heard from researchers around the world who are unravelling the genetic code to reveal if there are patterns of faulty genes in patients, or if there are ‘protective’ genes that stop someone developing the condition even if they have a faulty gene.

“John Hardy from UCL explained genetic research is already revealing how the conditions might arise. Most genes already found in patients with fronto-temporal dementia, PSP & CBD point to a problem with how the cell processes faulty tau protein – it cannot clear it away so it builds up. But he said we need more pieces of the jigsaw, and one way to do that is to do studies called ‘genome-wide association studies’ on samples from patients – where scientists do a ‘fishing exercise’ and screen the entire genetic code to look for faulty genes. He hopes that understanding PSP & CBD genetics better could give us more clues to new treatments.

“It’s not just genes in tau that could be affected – we heard about faults in genes and other molecules which control tau. And in PSP, different factors such as genes and environmental factors can affect how other genes behave - this fascinating subject is called epigenetics.

“Prof Huw Morris from UCL thinks some of the differences he sees in the clinic between patients with the same condition could be explained by genetics. In the Prospect study, they want to understand why certain patients get certain characteristics in their disease that someone else doesn’t. They are comparing people with PSP but different characteristics and looking for gene changes.

STOPPING FAULTY TAU SPREADING

“One of the highlights of the day for me was a talk by Dr Karen Duff from Columbia University in New York. She is interested in finding out how this faulty tau travels between cells in the brain across the gap – or synapse – between them, causing the disease to progress.

“Studying neurological diseases in detail is difficult in people, so Dr Duff’s team have developed a mouse that develops Alzheimer’s disease (AD), which is also caused by faulty tau. This mouse has revealed that faulty tau only affects certain types of brain nerve cells called excitatory cells – because they may be more vulnerable to attack. She’s then studied human brains with AD and brains from a patient with fronto-temporal dementia and found the same. This raised more questions. Why are only these cells affected - is it down to genes, or is it because these cells cannot control the amount of faulty tau that builds up? Dr Duff has discovered it’s the latter - these vulnerable nerve cells have higher levels of proteins in them that cause other proteins – such as tau - to build up. And these levels were higher in the regions of the brain that were affected in the disease first. This exciting work is revealing how these diseases are developing – and a way we could ultimately treat it using drugs.


MARIA TENNANT

“In the lab, the team have developed a fascinating way of studying how tau moves between individual cells. They are now using it to test if certain molecules can stop this happening. They are also working out the molecular process underpinning tau build-up – to see if they can prevent it. This research is an example of why understanding a disease properly is so important – it helps us to be clever about ways to treat it.

SOME EXCITING TRIALS

“The afternoon session was all about clinical research, where researchers are taking these lab findings and trying to turn them into treatments that would make a difference to patients. Because PSP & CBD are so rare, researchers are having to approach clinical trials differently, to make them more efficient. In the same trial, they test treatments on patients with different conditions but with the same underlying faulty genes, such as anti-tau antibodies which are being tested on both PSP & CBD patients.

“Researchers are also developing a clinical scale that doctors can use to monitor CBD, which could shape trials to make sure they are measuring the things that make a difference to patients. We heard updates about three anti-tau antibody treatments in, or about to start phase 2 clinical trials in PSP patients. The question obviously in my mind was around CBD - when a trial will happen in these patients. The answer was not yet, but the researchers hope this will happen soon.

THE DIFFICULTY OF DIAGNOSIS, AND THE VALUE OF PATIENT REGISTRIES

"I learnt it's incredibly difficult to diagnose CBD. Both PSP & CBD are ultimately confirmed by pathologists when they study the brain at post-mortem. Although PSP diagnoses are generally good and match the doctors' diagnosis, it's not the same story for CBD, and often at post mortem doctors find it was something else.

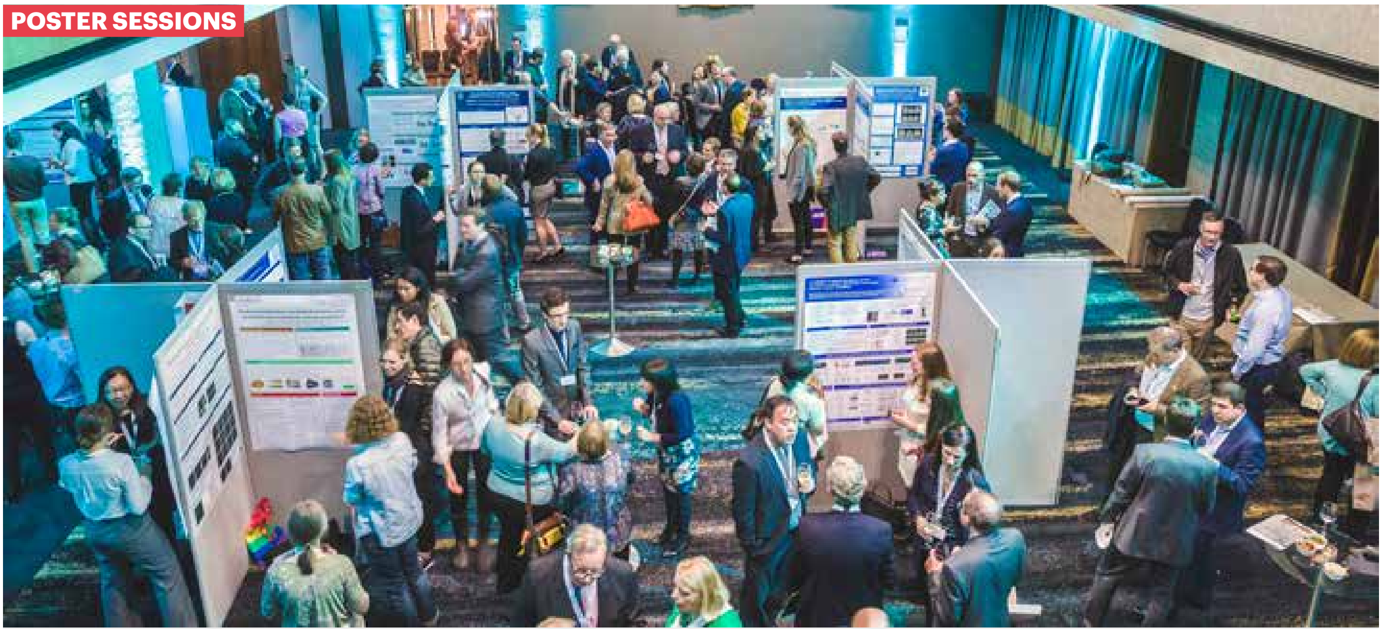
"This struck a chord with me. It has been a very long road to diagnosis with mum, probably around four years. Hearing this has helped to explain why this has been so long, because the diagnosis is so difficult for doctors to make. And it's even harder as people get older as other medical conditions can blur the lines between them.

"Doctors often want to test people earlier in their condition in trials, but this is difficult when diagnosis is later and can progress quite rapidly – there is only a small window to do it.

"So, what is the answer? Scientists believe it is monitoring patients for the long term. It can make diagnosis so much more accurate as we'll learn more about the conditions. Large patient registries where researchers build databases of patients with particular conditions allow doctors to collect a lot of information about patients as they live with their disease. They can reveal more about people with the condition, about the impact it has, and depending on the tests that are done, provide vital patient data such as genetics and brain scans.

"If those patients donate their brain to research, the scientists will have a full picture of how their disease has developed and how this has affected the brain itself. John Woodside from UCL gave an update on the Prospect study, which aims to make diagnosis more accurate and earlier, to help doctors understand disease types, find ways to predict disease progression, and to develop a PSP/CBD biobank. Having this database means they also have a 'trial ready cohort' – a group of people ready to go

POSTER SESSIONS



on a trial when treatments and funding are available. Mum has signed up to this, and we are hoping to get involved in the near future.

DON'T FORGET ABOUT BIOMARKERS

"The final session of the day covered biomarkers – ways to spot the disease and monitor its progression. This could either be a test such as a blood test, or sophisticated brain scans.

"James Rowe from the University of Cambridge told us about his exciting research bridging the gap between the lab and the clinic in Tau-PET imaging – where they can measure tau and work out where it is in the brain using brain scans. We also heard about research looking for biomarkers that act as 'molecular flags' and spot disease or measure its severity in a bodily fluid such as cerebrospinal fluid, or CSF, the fluid that surrounds the brain and spinal cord. We also heard about how MRI scans could detect and monitor these diseases, as it matches clinical decline.

REFLECTING ON THE CONFERENCE

"I now feel so much more knowledgeable than I did before – I understand what is happening within cells and where we are at with potential clinical trials. I feel like I can have a more educated conversation with my mum's doctors about the way forward and what she could get involved with.

"I won't lie, I wish CBD trials were a little closer than they are. But I will be watching out for developments in PSP and CBD research more closely and looking for opportunities that could help Mum and help to shape the future for others with CBD.

HELEN LING, UNIVERSITY COLLEGE LONDON

A MESSAGE FROM OUR SARA KOE PSPA RESEARCH FELLOW, ED JABBARI:

Dear PSPA supporters,
What a fantastic event the symposium was! It highlighted just how far we've come in terms of understanding the cause of conditions such as PSP & CBD, and what avenues we need to explore to develop effective treatments.

We had talks from drug companies about ongoing/upcoming clinical trials involving PSP patients. We're hoping to see the results of these trials in the next six months so watch this space. It was also great to hear talks on the various research cohorts of PSP/CBD patients being collected all over the world, including our PROSPECT-UK study. These cohorts are vital to gaining new insights into these conditions so we thank all of our research participants for the time and effort that they put into our studies.

The first day was an excellent chance to hear from patients/carers, therapists and doctors who are on the front line in the battle against PSP & CBD.

I'm grateful to PSPA and CurePSP for the opportunity to give a talk on the first evening of the symposium. This allowed me to discuss the various research projects I've been involved with over the last two years in my position as a PSPA funded research fellow, particularly our recently published genetic findings (page 5).

Overall, I came away from the symposium feeling very inspired! I'm very fortunate to be part of the PSP research community at a time when there is so much momentum and optimism. We should all remain hopeful.

Ed

MUSICAL CONCERT ON FATHER'S DAY FOR PSPA

Actors Louise and Stephen Palfreman, pulled together some of the leading stars from the West End to put on a musical concert to raise money for PSPA. Louise talks to *PSP Matters* about her very personal reason for supporting the charity and how each member of her family has found their own way to help raise money to find a cure.

"The first thing was the random falls. Then the struggle to park his car. The changes were subtle, but gradually it was time for my 'doctor shy' Dad to make a GP appointment. My dad, Norman, a fit, active and intelligent man was unable to complete the simplest of tasks. Dad underwent many tests, all inconclusive. Knowing something was seriously wrong, we persevered with the GP. Eventually, after a two day stay in hospital, he was given the news he had PSP.

"Our relief at finally finding out what was wrong soon turned to fear. Over the coming days it dawned on us, Dad had an incurable brain disease. Incurable – the word you never want to hear said about a loved one. Dad, typically for him, remains positive. He is currently on a clinical trial and is determined to get to the clinic no matter what. Recently he had a horrific fall, but still the following week he was back at the clinic for his next infusion. It's not just Dad living with the impact of PSP, but my mum too. Mum now has to do everything, so it has been a steep learning curve for her. Watching her cut up wood for the log burner, mow the grass and do all the things Dad used to do brings it home how much they have both lost. It also brings it home what a great team they are and how much they love each other.

"We are a close family and each of us separately found ways to deal with Dad's PSP. There was no big family discussion, but for each of us doing something positive felt like the only option. We all wanted to do something to show Dad how much we love him. When you are in such a hopeless situation, it makes you strive to do something where some good will come of it.

"My brother, sister and I have each chosen to support PSPA in ways that play to our own strengths. My brother and sister are keen runners and each took part in running events. Not fond of running, I chose to use my work as an actor in the West End to raise funds. Along with my husband Stephen, also a West End actor, we decided to put on a musical concert for PSPA.

"We approached leading West End producer Bill Kenwright who owns the Theatre Royal in Windsor and asked if we could book the theatre on Father's Day, as a tribute to my dad. Naturally I aimed to raise as much money as possible, but I also wanted to raise awareness. No one has heard of PSP, and that is very frustrating. As soon as we started pulling together the programme we had so much interest, with approaches from radio shows and newspapers.

"Stephen and I called upon all our friends and colleagues in the West End to help. We pulled together a brilliant cast with some leading names, including TV actors Neil McDermott from EastEnders, and Amy Robbins and her husband Robert Daws from The Royal.

"I was touched everyone came along to support us. Our thank you list was certainly a long one. Often, in times like this you get to see how kind people really are. From Dad's previous employers to local businesses, everyone did something to help. We raised £5,400, including £1,400 that was raised by a raffle at Mum's workplace, Nationwide Building Society.

"At the end of the concert Dad got on stage to say a few words. It brought a tear to everyone's eyes. He was clearly moved, as we all were. It was a wonderful way to show Dad how much he is loved by so many people. Fundraising for us is a way to show Dad how much he means to us. When something like PSP comes into your life, you feel so helpless. In a situation where you're told there is no cure, trying to find something positive to focus on helps.

LOUISE'S MUM RAISES MONEY AT WORK

LOUISE AND HER DAD NORMAN

"The Debbie McGee show on BBC Radio Berkshire want us to come back once Dad has finished the clinical trial. So, long after the curtains closed on our concert, we will continue to raise awareness of PSP, and money so we will one day find a cure to beat this devastating disease."

"FUNDRAISING FOR US IS A WAY TO SHOW DAD HOW MUCH HE MEANS TO US."

"WHATEVER I'LL DO, IT WILL BE FOR PSPA."

Melanie Duffin's stepfather Ray died from PSP. Melanie shares how her personal experience of seeing the devastation caused by PSP drove her to take on a stage of the Tour de France.

"It was the small things my mum, Carol noticed at first. She is a retired nurse and recognised early on that something was not right with my stepfather, Ray. He had many visits to the doctor with a range of symptoms varying from visual problems, falling over backwards, and severe neck pain, yet nothing was ever resolved. Many of the medical professionals he encountered during this time had never even heard of the disease, so recognising it and dealing with the issues was difficult.

"Mum and Ray went through months of trying to find out what was wrong, though not having a diagnosis was so difficult for them. My mum would find herself becoming frustrated with Ray for making silly errors. This was a saddening and confusing time for both of them.

"For me an early diagnosis and a recognition of the early signs is vital. Ray had continued to drive but noticed himself his reactions were too slow, and he was having little accidents. Luckily nothing major as he was driving around his grandchildren. This could have ended very differently due to not knowing what was wrong.

"Mum and Ray came to visit me in France, where I was living at the time. We found Ray was unable to cycle a bike without falling off, and could no longer manage to swim in the pool, his co-ordination was lost, and his fixed stare was getting worse. That was the point Mum knew it was something serious. For a man who was very active and loved to be outdoors doing many activities, the harsh reality was dawning on mum that she was losing her husband. Mum had read enough to realise that it might be PSP. She asked the doctor and he confirmed she was correct.

"Every time I came home, I'd see a big deterioration in Ray's condition. The changes were so rapid. Mum was determined to give him the best quality of life until the very end. It was always his dream to travel around Scotland in a camper van, so they bought a blue one, and travelled for 120 days. This gave Ray his dignity while travelling. They enjoyed listening to the birds singing and their favourite music, one being Dignity by Deacon Blue, hence the van being named "Bluey".

"I wanted to do something to raise awareness and help fund research. I decided to do a stage of the Tour de France, however was very aware I wasn't strong enough. It is not a race people usually do for charity, but I wanted to use it as a personal challenge and a way to raise money for PSPA. The race was 169km and involved a 4,070 metre climb – it was hard and took a lot of training. Finally, I completed it in 8 hours and 59 minutes… not quite 9 hours!

"Sadly, Ray died at home with Mum by his side, so was never able to see what I did, but he knew what I was doing as Mum kept him updated. Though he couldn't see or move much he was still aware of his surroundings, and what was happening to him. Later in the year, I moved back to England for many reasons and one of them was to be closer to mum. We have done quite a bit of fundraising since. It has helped us to focus on doing something positive.

"Now, I need to find my next goal. I have a few ideas in mind, but nothing confirmed yet. However, whatever I do it will be for PSPA."

WHAT COULD YOU DO IN 2019?

2019 STARTS HERE – WHAT CHALLENGE COULD YOU DO?

Many of us set new year's resolutions to get fitter or take on a personal challenge. Perhaps one of the exciting events below may be just what you need as we enter into 2019.

RIDELONDON – 4 AUGUST

100 miles of cycling glory await! From the iconic Olympic Park, through Surrey's green and pleasant country, to the triumphant finish along The Mall, RideLondon is the UK's biggest and best cycling event. Join PSPA's biggest-ever team and make history!

BRITISH 10K – 14 JULY

A 10K run through the heart of London, taking in all of its world-famous landmarks. The British 10K is such a fun event, and you can join Team PSPA in 2019.

ROYAL PARKS HALF MARATHON – 13 OCTOBER

A huge event on the running calendar! Nothing beats running through the array of Autumnal colours of London's Royal Parks!

GREAT NORTH RUN – 8 SEPTEMBER

The most famous half marathon in the world, you can be part of something truly special by running this wonderful event for Team PSPA.

OVERSEAS ADVENTURES – ALL YEAR ROUND

The world is waiting to be explored, and it waits for you. Do something that will live long in the memory and take on one of our epic overseas challenges.

- See the natural wonder of Iceland on our Iceland Highland Trek.
- Cycle through steamy jungle and sleepy villages on our Vietnam to Cambodia cycle.
- Trek to the Roof of Africa on our once in a lifetime Kilimanjaro challenge.
- Wonder at the beauty of the Andes as you climb to Machu Picchu on our incredible Inca Trek.
- Explore secret Burma on two wheels on our Road to Mandalay adventure.
- Trek along the Great Wall of China and seek out the ancient watchtowers.

DENISE HUNT AND HER MUM

THE FRONT LINE IN AWARENESS RAISING AND EDUCATION

Last year we created a new volunteering role to help educate health and social care professionals about PSP & CBD. We know the difference it makes to have health professionals who are aware of PSP & CBD and how it impacts people and their families. That is why we are determined to focus our efforts on educating those best placed to make a difference to people living with PSP & CBD.

We now have a team of education volunteers who are focused on raising awareness in their areas. Most have personally seen the devastation caused by PSP & CBD and wanted to do something to help others affected. *PSP Matters* spoke to Denise Hunt and Charlotte Harmon about their motivations for volunteering and what they hope to achieve.

Police officer Charlotte Harmon's grandad, Bernard was diagnosed with PSP four years ago and sadly died in 2018. Charlotte said: "I had never considered volunteering until I saw an appeal for more volunteers on PSPA's Facebook page. I have previously done quite a bit of fundraising for PSPA, including running the London Marathon, but this year I thought volunteering may provide a great way to support the charity and gives my friends and family a break from me asking for donations!

"I am so grateful for everything PSPA has done for my nanna and grandad, so volunteering is a good way for me to show this. When I heard about the educational volunteer role I knew it was perfect for me. One of our biggest frustrations is that no one has heard about PSP. Now I can do something constructive to help. As a police officer I do shift work. Volunteering fits around my full-time role easily and work is very supportive.

"I completed the online training course. Now I am getting my presentation together and in the coming weeks I will be contacting care homes and GP surgeries to ask if I can give a talk. I do feel a bit nervous, but I'm also excited to be part of the effort to educate more health and social care professionals about PSP & CBD. The more they know about it the better care they can give people.

"My nana, Janet set up the Local Support Group in Lincolnshire. She saw there was no local support around Lincoln, so decided to set up a group herself. I am so proud of her. I'd like to follow the example she has set in helping others.

"As a family we have always been strong. My grandad helped to raise me and has always been my role model.

"I'm excited about focusing on my new education role in 2019. My number one message to people will be, 'it's not Parkinson's'. Misdiagnosis is horrible and unfortunately so many of us have had to go through it. We had years of not knowing what was wrong. Diagnosis has to be improved. It is unfair to leave families living with PSP & CBD without a diagnosis for so long."

Denise Hunt's mum was diagnosed with PSP after four years of tests and appointments. Denise wants to help improve professional's knowledge so that diagnosis will not take so long for others.

"Mum has suffered a great deal, not just physically, but mentally too. She has endured the frustration of thinking no one believed her, until eventually, after four years of appointments a diagnosis of PSP was confirmed. This long and exhausting journey to diagnosis is what drove me to become a PSPA education volunteer. I am determined to help increase knowledge and awareness of PSP & CBD, to save other families from what we went through.

"I want families to receive an earlier diagnosis, so they can then focus on making the most of the time they have left. I want families to spend more time together making memories and less time in hospital waiting rooms.

"The impact of not getting a diagnosis is so far reaching. I would get cross with Mum, especially as she appeared to get more and more obsessed with feeling unwell. Now, with hindsight, I realise she was worried, and, was also undergoing changes to her mood and personality.

"You can't cure PSP, but if more was known about the condition we wouldn't have wasted so much time going back and forth to various consultants. Also, we would have been kinder and more understanding towards what Mum was going through. Once we had received the diagnosis, everything fell into place.

"After Mum's diagnosis I was reading PSPA's website. I would absorb all the information, printing off the leaflets and resources for healthcare professionals. I saw PSPA was appealing for more volunteers. When I saw the educational role, I knew that would fit well with my experience working in the pharmaceutical industry where I would often talk to healthcare professionals.

"I want to help improve diagnosis. If I can save just one person from the endless string of appointments that would be the biggest gift ever. Since completing my training as an education volunteer I have contacted a number of organisations to ask if I could come and give a talk. I can fit my volunteering around my work, and it helps that the organisation I work for is very supportive of what I want to do. My drive to educate people about PSP & CBD keeps me focused on doing something positive in the face of the most devastating news I could ever have heard about my beloved Mum."

IF I CAN SAVE JUST ONE PERSON FROM THE ENDLESS STRING OF APPOINTMENTS THAT WOULD BE THE BIGGEST GIFT EVER.

If you would like to become an education volunteer and raise awareness amongst health professionals please contact **volunteering@pspassociation.org.uk** or **01327 356144**.

CHARLOTTE AND HER GRANDAD

CBD MATTERS

NEW PSPA *STARTS HERE*

LISA RODRIGUEZ WHO IS
LIVING WITH CBD OPENS
OUR NEW OFFICES

CONTENTS

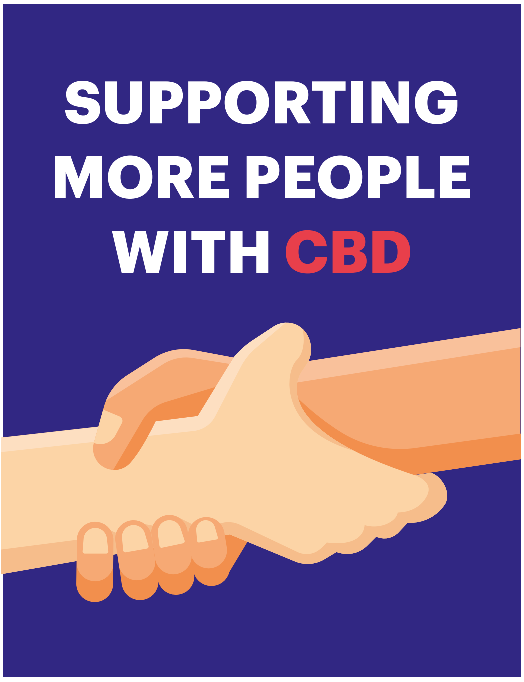
CBD WELCOME

Welcome to *CBD Matters*. We are determined to produce more CBD-specific information and support more people affected by CBD. Part of our plans to do this includes the launch of our Professional's Guide to CBD. We hope this important resource will enable health care professionals to provide the very best care to people with CBD, their carers and families. To mark the publication of this important resource, we decided to focus on CBD in this bumper edition of *PSP Matters*. Alongside this, we are currently producing information for people living with CBD and their families that will be available from the spring.

Thank you for all your continued support and I wish you a very happy and peaceful New Year.

Andrew Symons
Chief Executive

CBD NEWS



WE NOW SUPPORT MORE PEOPLE WITH CBD

With CBD & PSP sharing some similar traits and symptoms, and the fact that people with CBD were left to cope alone, with no charity to support them, the decision was made some years ago to extend our charitable activities to cover CBD. In 2011 we had three people with CBD registered with us. This number has increased to 281 people living with CBD and their families. If you know someone living with CBD who is not in touch with us, please do share this magazine with them - we are keen to reach as many people as possible affected by CBD.

PROFESSIONAL'S GUIDE TO CBD

Since the launch in October, we have sent out nearly 500 copies of the guide to health and social care professionals.

NEW OFFICE OPENING

Lisa Rodriguez 48, who, last year was diagnosed with CBD opened our new offices in Milton Keynes. Lisa was joined by local supporters and health and social care professionals to mark the occasions. PSPA Founder Michael Koe also came along to the event. Speaking to media at the event Lisa Rodriguez said: "I know CBD is life limiting and I will progressively get worse. All I can do now is stay positive and make the most of my life. I am also determined to do as much as I can to raise awareness of CBD and help push forward research into the condition. Having the national charity now on my doorstep makes me even more focused to do as much as I can."

RED FLAGS FOR CBD

We have launched our Red Flags for CBD. We know GPs often find it difficult to spot symptoms of CBD and that many may not have come across the disease before. Sadly, this often means people receive a delayed diagnosis, or are misdiagnosed with other conditions. So we have developed a Red Flag document you can share with your GP practice as well as other health professionals you come into contact with. They in turn can share it with their teams and raise awareness of CBD. If you would like some Red Flags to hand out please contact PSPA Helpline.

COULD YOU MAKE A REGULAR GIFT IN OUR 25TH ANNIVERSARY YEAR?

This year is a special year for PSPA. It is our 25th anniversary and we plan to use this year to drive forward improvements in how we support people with PSP & CBD by raising more money and recruiting more education and support worker volunteers. However to do that we need your help.

Regular donations make a huge difference to our work in funding research into PSP & CBD, and providing information and support to all those affected by both devastating conditions. By choosing to give a regular monthly donation, you will be helping us to plan ahead and commit to funding research projects. A regular gift allows us to plan for the future because we know, day in day out, that we will have a steady income stream we can rely upon.

This birthday year, would you consider making a regular gift of £5, £10, or £25 to help secure our future work? You can make a gift here **www.pspassociation.org.uk/fundraising/donate/give-now-or-regularly/** or contact our friendly fundraising team on **01327 356132**.

“I ALWAYS WANT TO LOOK AFTER MUM.”

Claire Wells is a carer for her mum Anna, who is living with CBD. Claire shares her experiences of being a young carer and how CBD has affected them both.

“Life has changed for us considerably since Mum was diagnosed with CBD. Mum had to leave her two bed Victorian home in Surrey for a more accessible house. Her partner of ten years couldn’t deal with her progressive disability and decided ‘he wanted his life back’ so left her. At the time I was working in London. I altered my working hours so I could support Mum more. Eventually, we decided to sell the house and relocate to Milton Keynes, where I had some close friends.

“We found a house, but it needed a lot of work to make it suitable for Mum. I took a two-month sabbatical from work to focus on getting the house ready, complete with ramps and all the things you don’t ever think about when you are able-bodied. At the end of my sabbatical it wasn’t feasible for me to return to my job in London. Of course, I miss a lot of the perks of working, but my priorities have changed hugely.

“In April, Mum had a nasty fall in the shower and fractured her spine. She has been in a wheelchair ever since. That is the point we decided to get carers in to help, despite her initial concerns about strangers coming in to look after her.

“Mum naturally wanted to keep doing as much as possible, but I wish we had known more about the different stages of the condition. My advice to anyone now would be not to resist using a wheelchair and not to think about it as the end of the road. Looking back, had Mum used a wheelchair just when she needed to, I think it would have saved her from being in a wheelchair permanently.

“It was only in January we received a diagnosis of CBD, after 11 years of Mum experiencing symptoms. She would be constantly told she was making it up. Her GP described her as ‘anxious’. A surgeon put her symptoms down to her not wanting to accept that she’s simply getting old. Different doctors told Mum different things, including at one point a diagnosis of Parkinson’s.

“It is quite shocking how Mum deteriorates from one week to the next. Her memory is still good but she sleeps 60 per cent of the day and is often confused when she wakes up. She'd ask when Dad is coming home, but he died when I was 21. The change in personality and temperament is the hardest thing to deal with, it really upsets me. In a way I prefer helping her physically, because the mental side is very difficult, she can get very down.

"We have had so many issues with carers. Most have just not understood her condition and why she can't move her leg. I think one of the challenges is because the care industry is so badly paid no one thinks of it as a career. Many carers don't understand Mum has a neurological condition. We did have one wonderful carer, but she left to become a paramedic. It made such a difference to have someone who got on well with Mum and who I could rely on.

"At the moment I am doing 99 per cent of the care and that can be quite stressful. I get Mum out of bed and transfer her onto the shower commode. It takes about an hour to get her washed and dressed. The night times are getting harder. I don't know if it is a side effect of the new medication she is on, but she needs to go to the toilet at least six times during the night. It is exhausting for the both of us. If I do go out we need to get a waking night carer at £230 minimum to sit downstairs for when Mum needs help.

"I do feel selfish sometimes for wanting a life, but to have a life and do things I enjoy we need to have carers we can rely on. This is something we're still working on. Mum feels guilty I am caring for her. I feel guilty about wanting to go out sometimes. But, at the same time, I always want to look after Mum. When we have good carers again, we'll be able to go back to having a typical mother/daughter relationship, but at the moment in the absence of extra support, I am her main carer, best friend and daughter.

"We have always both been keen travellers and I have been adamant that we try and maintain a good quality of life. We went to Amsterdam in September on the direct new Eurostar route. Accessible Netherlands arranged the equipment we needed for the room and it ended up being a home from home. We try to do as much as we can. Mum has had a flying lesson and went sailing. We had a lovely day out recently where we fed the penguins. Mum has always found it hilarious that when I was younger I called them pupuins. It's important we have fun and focus on what Mum can do and enjoy.

"For years we felt lost in the system with no one to turn to for information. That changed once Mum was diagnosed. That evening we found PSPA online. They put us in touch with our local Neurological Nurse Practitioner who has been a huge help and given us names of people to help with adaptations, equipment and other professionals for support.

"CBD has changed Mum. She was so bubbly and outgoing, always turning heads. She loved life, was always off climbing mountains and swimming in lakes. I know she will never come to terms with the fact that she can't do those things anymore. It's frustrating to watch how she is treated by people who didn't know her previously. She's only 67 but she's often patronised and spoken to like she's way older, or worse still, they will speak to me instead.

"It is very lonely to be an only child with no family to rely on for support. However, I am just happy I am in this area and I have close friends nearby. Through my Mum's experience I really value the importance of friendships.

"Despite everything that has happened I am happy. I have discovered a lot about myself and what I truly want to do with my life. Of course my life is very different to when I was working in London, but the London scene doesn't appeal to me as much anymore. Now I know I would like a caring role, or perhaps work for a charity. The situation I am in has changed me for the better. I have also met some fantastic people and I'm so much more appreciative of my own physical health and wellbeing.

"I started running and did the couch to 5K. I'm now training for the London Marathon next April for PSPA. Another thing I perhaps wouldn't have done if my life had been different."

CLAIRE AND HER MUM FEEDING THE PENGUINS

THE WORLD'S FIRST PROFESSIONAL GUIDE ON CBD

PSPA has produced the world's first professional guide on CBD. The guide is designed to be the 'gold-standard' for professionals planning and delivering care to people living with CBD.

A Professional's Guide to Corticobasal Degeneration was launched to health and social care professionals at our Study Day event last October. Funded by CBD Solutions, the development of this guide brought together health and social care professionals and people living with CBD, their carers and family members.

We know that a lack of knowledge and awareness of CBD can hamper health professionals' ability to provide good care. Alongside this, issues around care co-ordination can also mean families are having to deal with a large number of professionals who may be unfamiliar with issues faced by the person living with CBD. Many health professionals have commented on the impact this guide will have. Described by Prof David Oliver, Consultant in Palliative Medicine as 'an exciting development' our guide helps to address these issues by providing a blue print for the integration of care by setting out a framework for care delivery.

Prof David Oliver said: "This guide provides vital information to health and social care professionals who may have little experience with CBD patients. It details good information and advice to help improve the care of people with CBD, from diagnosis to the end of life."

This new guide provides an overview of the standards of care for people with CBD, their carers and family members should expect. It will be complemented by an on-line interactive resource and up-to-date framework that will be able to be viewed on a desktop, tablet or mobile phone. It has been written specifically for health and social care professionals, but we are currently working on a version specially for people living with CBD, their carers and families which will be available from April 2019.

Sarah Agyekum is a GP and was part of the working group to help develop the guide. Sarah said: "I was delighted to be invited onto the working group for the Professional's Guide to CBD. We all remember as young GPs that we were taught the mantra 'common things are common' when dealing with diagnostic uncertainty.

"However, it was often those much rarer conditions like CBD that we worried about missing. Clearly the diagnosis of conditions like CBD remain challenging, particularly to the non-specialist, as so often in the early stages they share overlapping features with more common conditions such as Parkinson's.

"This guide not only educates GPs to be on the look-out for early hallmark signs (red flags) that can help us consider CBD as a potential diagnosis much earlier on, it also helps us to support people once diagnosed with the condition with a clear concise guide to symptom management and expected standards of care. Central to this is the role of the multidisciplinary team and the importance of advance care planning, providing holistic care and support to patients and their carers throughout the course of the disease. I look forward to sharing the interactive web-based resource with my colleagues and spreading the word about CBD."

AVAILABLE NOW

A Professional's Guide to Corticobasal Degeneration is available to download **www.pspassociation.org.uk/app/uploads/2018/11/PSPA-CBD-Guide-A5-LR.pdf** It can also be ordered from our helpline **0300 0110 122 – helpline@pspassociation.org.uk**

"MY PATIENTS' ATTITUDE AND DETERMINATION TO HELP OTHERS MOTIVATES ME EVERY DAY TO DO WHAT I CAN TO IMPROVE THE DIAGNOSIS OF CBD."

Dr Ruth Lamb at the UCL Queen Square Institute of Neurology describes her work on setting up a European registry of CBD patients and how researchers are seeking to improve diagnosis.

"My clinical research post, funded by CBD Solutions, is to set up a European registry of CBD patients. Alongside this, we are linking together movement disorder clinics in hospitals across Europe, who have an interest in and are able to recruit people living with CBD. This will enable study sites to be set up in key hospitals, where clinicians can undertake research projects and recruit patients for future trials. My research post also oversees the UK recruitment of CBD patients as part of the PROSPECT study, headed up by Prof Morris.

"Now, three years into my project, we currently have 165 patients with CBD syndrome recruited to the largest ever study of patients with CBD in the world. CBD is rarer than PSP and is harder to diagnose, with features far more variable than PSP. A person with CBD may not have all of the features we classically see in CBD and sometimes CBD is mimicked by other conditions such as Alzheimer's, PSP or frontal temporal dementia. We tend to describe patients as having a CBD syndrome, rather than CBD. That is because currently, we can only truly diagnose people with CBD after they have died, when we are able to look at their brain under a microscope.

“We have looked at the accuracy rates for diagnosing CBD, by studying the brains of people previously diagnosed after they had died. We have confirmed we are not very good at diagnosing people, with around half of people diagnosed incorrectly. Symptoms can progress quite slowly over time, and it may take some time for the syndrome and the diagnosis to become apparent. Another challenge is most clinicians will never see a patient with CBD. People, including health professionals, are not aware CBD exists. That is why it is really important to raise awareness of CBD, how it presents itself and the importance of a speedy referral to a neurologist or movement disorders specialist.

“Often by the time patients are diagnosed they have spent many months or years going around the health system. We think this relates in part to subtle and non-specific symptoms early in the disease course (eg clumsiness of one hand, subtle changes in balance or personality) and in part to lack of awareness of the condition.

“We are investigating what kind of tests can help us improve diagnosis, like MRI scans, biomarkers and cognitive tests. Patients naturally want a diagnosis and not having one when something is clearly wrong can be difficult. It can also make it difficult to explain to their friends and family what is going on. Without an accurate diagnosis, patients most likely will be deprived of accessing the right type of support from health and social care professionals. Ensuring patients receive a correct diagnosis is also vital for the future, for developing and testing new potential treatments for CBD.

“In conversations with people who have CBD syndrome it is clear many feel isolated. No one knows what the condition is or its impact on them and their family. With such low awareness they are having to become specialists in the condition themselves. They find it exhausting having to explain CBD to every professional they see and find the added stress really overwhelming.

“We think some of the tests we are currently investigating will be helpful in predicting an underlying diagnosis of CBD. These patients are the ones we would want to put forward to clinical trials targeting the tau protein.

“Patients are naturally very interested in research and many are keen to be involved, even if they know it won’t help them. Our patients say that research gives them hope and it provides them with an opportunity to turn their experience into something positive that will help others. Their attitude and determination to help others motivates me every day to do whatever I can to improve the diagnosis of CBD. By doing that, we will in turn be improving the care and, importantly, pushing forward the discovery of future treatments to slow or halt the progression of CBD.”

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If you would like to fund research into CBD please contact **info@pspassociation.org.uk**