



RARE DEMENTIA SUPPORT

PPA Support Group Newsletter – August 2018

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Welcome

Welcome to the August 2018 edition of the PPA Support Group newsletter. We aim to give you some insight into PPA research around the world, and information about our upcoming meeting.

We know that we cannot possibly imagine what it must be like to have a diagnosis of PPA, or to know and love somebody who has the diagnosis. We would like these newsletters, and indeed our Support Group meetings, to be shaped by what you would find most useful so please do get in touch with Chris Hardy by email at c.hardy.12@ucl.ac.uk or leave a message on 07388 220 355 if you have any suggestions, or if you would like to talk to him about PPA and how it is affecting you or somebody else.

In this newsletter, you can read a brief summary of our April meeting (page 2), meet some new members of the team (page 3), and hear about some of the latest research to come out of UCL and internationally (pages 4 and 5).

We have an update from the National Brain Appeal on page 7, and this is where you might want to start if you have only recently heard about PPA – we have included a brief description there.

Our next meeting will take place on Thursday 6th September 2018 and we have taken on some feedback and decided to trial a new format for that meeting – you can read more about this on page 6.

We would also like to bring your attention to our new website (see photograph below). Our team here has been working very hard behind the scenes to improve our previous offering, and we hope that you will like our new site: www.raredementiasupport.org/ppa

Please do let us know if you have any feedback on how we might improve it still further.

Yours faithfully,

The PPA Support Group Team

1. Welcome



Our last meeting

It was a great pleasure to see so many of you at our last meeting in April. In the morning, we heard from **Dr Thomas Cope** from Cambridge University. Thomas presented some of his fascinating research on the nonfluent variant of primary progressive aphasia. His research attempts to explain why people with nfvPPA can have difficulties understanding speech, even in quiet situations. In healthy people, the frontal 'speech' parts of the brain constantly make predictions about what somebody is saying – and those predictions help the temporal 'hearing' parts of the brain to understand what another person is saying. These predictions have to be continually and constantly updated. When these frontal regions are damaged in the course of nfvPPA, the person can start making what Thomas referred to as 'inflexible predictions' – which might explain why people with nfvPPA have problems understanding speech.

After lunch, we heard from **Anna Volkmer**, a speech and language therapist from UCL. Anna gave an overview of some work that she has been doing during her PhD to develop an online therapy resource to support speech and language therapists to deliver communication skills training to people with PPA and their families. In the latter part of her presentation, Anna focussed on 'hints and tips' for communication in PPA – which was a really interesting and helpful way to end the day.

11th International Conference on Frontotemporal Dementias (ICFTD)

We also wanted to let you know that the 11th International Conference on frontotemporal dementia will take place in Sydney in November of this year. This is the major (biennial) conference dedicated to FTD and PPA and helps set the research agenda and clinical practice standards on a global stage.

Researchers from UCL will be well-represented at the conference – and in our next newsletter we will update you on the work presented there.



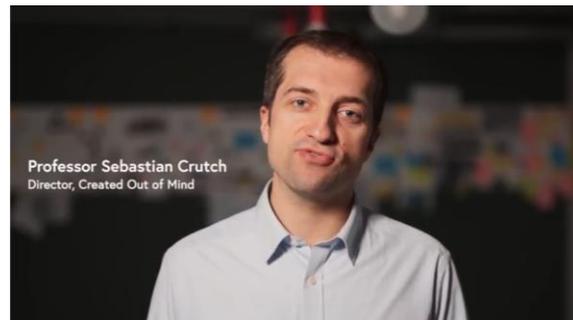
Tell your story of living with primary progressive aphasia

Would you like to share your experiences?

We are working in collaboration with the Alzheimer's Society to raise awareness of the experiences of people living with the more rare forms of dementia, including PPA. By talking about what you've been going through, you can help Alzheimer's Society tell powerful stories that engage hearts and minds and ensure all types of dementia are a public and political health priority. Whether you are living with PPA or caring for someone with the condition we'd love to hear from you. By raising awareness, experiences like yours can change attitudes, challenge stigma and help fund vital research.

If you'd like to find out more please contact: stories@alzheimers.org.uk, or email Chris at c.hardy.12@ucl.ac.uk who will be happy to talk to you about your involvement.

Massive Open Online Course – Dementia and the Arts: Sharing Practice, Developing Understanding and Enhancing Lives



The Dementia Research Centre and Created Out of Mind have launched a new four-week online course exploring how the arts can create a common ground between people, and in doing so, learn what we can all do to improve the quality of life and care for people living with different dementias. Topics covered include:

- Changing Perceptions: Can the arts help us see the dementias differently, and can the dementias help us see the arts differently?
- Creating Common Ground: The arts, equality and dementia

Please refer to the website if you are interested in finding out more information: <https://www.futurelearn.com/courses/dementia-arts>. It's completely free to join, and sign up is open now with the first run due to begin on 10th September 2018.

Saying hello

Dr Kirsten Perkins has joined the Rare Dementia Support team, and is also part of UCLH and based at the Dementia Research Centre. Kirsten is a chartered Clinical Psychologist with the British Psychological Society (BPS) and Associate Fellow of the BPS Division of Neuropsychology. She has experience in developing neuropsychology diagnostic and rehabilitation services in a number of different areas, including in-patient, out-patient and community settings. She has worked with a wide range of people with neurological disorders including dementia, Parkinson's disease, multiple sclerosis, stroke, and epilepsy. In her role at the DRC, she will be providing psychological assessment and intervention to people presenting with a range of psychological, behavioural and cognitive difficulties. She also provides psychological support to relatives and carers.

We would also like to take this opportunity to introduce you to **Roberta McKee-Jackson**. Roberta has been involved with the posterior cortical atrophy (a type of dementia that affects visual processing) support group for many years and we are delighted that she – with her wealth of knowledge and experience of both the support groups and the rare dementia journey - has formally joined our team as Rare Dementia Support Regional Coordinator. Roberta will be supporting our team of Regional Facilitators in their running of support group meetings for all the types of rarer dementias that fall under the Rare Dementia Support umbrella.



**Roberta
McKee-Jackson**



**Kirsten
Perkins**

Saying goodbye

Sadly, we also have one 'goodbye' to say, to Dr Charles Marshall. Charlie has been a stalwart of the PPA Support Group for many years. Some of you will know Charlie clinically – he has worked as a neurologist at the National Hospital for Neurology and Neurosurgery whilst undertaking a PhD with Professor Jason Warren at the Dementia

Research Centre, UCL. Charlie has been extremely generous with his time at our support group meetings in the past: presenting, facilitating Q & A sessions, and generally being a happy and smiling face over coffee and lunch. Charlie has accepted a post as a Clinical Lecturer at Barts and the London. He says:

"It has been a great pleasure to get to know so many of you over the last four years. It has been an inspiration to see the proactive roles you have taken in the support group and in research in spite of some of the challenges that PPA presents. Together, we are constantly improving our understanding of PPA, including how to live well with it, and we will soon be planning treatment trials. I am not moving very far, and I hope that I will still remain connected to the Support Group to share in this progress. Thank you to all of you."

We wish him all the best – and think it quite likely that we will try to tempt him back to share his research with us in the future! Congratulations Charlie for an outstanding PhD, and thank you for your dedication and passion for research into FTD and PPA.



Charlie Marshall

Research Update

Something that we trialled in our last newsletter and that seemed quite popular was providing a research summary of the ground-breaking research taking part at UCL and around the world. Chris Hardy has offered to continue this, but again offers the following statement:

"I promise that I will try to pick papers that I think will be most interesting and relevant to our members – but I may not always get this right – so please do give feedback if I'm straying too far from what would be useful here. The idea is that although we have all of this incredible research going on into PPA, it is often written up in such a way that makes it really hard for non-scientists to understand – so hopefully I will be able to translate some of the scientific jargon into accessible summaries in these newsletters. I should also say that these summaries will be limited by my own understanding of the papers, and so apologies if I make any errors!"

1. Retained capacity for perceptual learning of degraded speech in primary progressive aphasia and Alzheimer's disease

Lead authors: Chris Hardy and Jason Warren
Where published: Alzheimer's Research and Therapy

When: August 2018

Summary: This is one of my own papers – so I hope that I can do it justice here! Most research in PPA tries to define 'deficits' – investigating what people with PPA can no longer do. There has been very little work done looking at brain 'plasticity' in PPA – this same plasticity is what allows people to recover well after having a stroke, for example. In this study, we wanted to investigate understanding of speech – something that people with PPA tell us gets harder – and that can cause significant social and communication barriers. We used speech that had been very 'distorted' – similarly to what happens in the real world where you have to focus in on a single person talking over a babble of background speech. To start with, people with the nonfluent and logopenic forms of PPA found it very difficult to understand this distorted speech – but over time they became much better. I think this is potentially really important – it provides evidence that behavioural treatment and rehabilitation strategies, like speech and language therapy, really do work in PPA.

2. Cardiac responses to viewing facial emotion differentiate frontotemporal dementia

Lead authors: Charles Marshall and Jason Warren

Where published: Annals of Clinical and Translational Neurology

When: April 2018

Summary: In this study, Charlie asked people with behavioural variant FTD, semantic variant PPA, and nonfluent variant PPA, to look at videos of facial expressions and to identify the emotion that the person was displaying, all whilst he was measuring their heart rates. He found that on average, all three participant groups had difficulties in accurately identifying the emotions. However, Charlie was also measuring the participants' heart rates – watching emotional stimuli produces heart rate changes in healthy individuals – and we know that these heart rate changes help us to (subconsciously) understand the emotions

that we see in others. Crucially, the participant groups differed when Charlie looked at the change in heart rate in response to the videos. In people with semantic variant PPA, heart rate changed on viewing the emotions – just as he saw when a group of healthy participants did the task as well. But in people with behavioural variant FTD and nonfluent variant PPA, this 'cardiac reactivity' was lower than in healthy people. These results are really interesting because they imply that the problems people with PPA and FTD have with recognising emotions are caused by different reasons. People with damage to the very front part of the temporal lobes (as in svPPA) are likely to lose their understanding of what each emotion is or means, which explains their difficulties on this task. However, people with nfvPPA might have problems because their body doesn't give them the same clues that healthy people get.

3. Thalamic atrophy in frontotemporal dementia – Not just a C9orf72 problem

Lead authors: Martina Bocchetta and Jonathan Rohrer

Where published: Neuroimage Clinical

When: February 2018

Summary: This work was led by Dr Martina Bocchetta, a postdoctoral researcher at UCL working with Dr Jonathan Rohrer. The thalamus is a small structure deep within the brain that transmits signals from our body to our brain so that we can understand them. It is also involved in managing our sleep, alertness and consciousness, and previous research had shown us that this region was particularly vulnerable in people with a genetic mutation in the C9orf72 gene – something that we know causes behavioural variant FTD in some people. Here, Martina looked at the brain scans of 341 different people with FTD, including those with semantic variant PPA, nonfluent variant PPA, and people who had been given a diagnosis of PPA that didn't fit into one of the three major subtypes. Martina found that, despite what had previously been thought, damage to the thalamus doesn't just occur in people with C9orf72 – it happens to all different kinds of FTD. All of this means that we could use the volume of the thalamus as a 'biomarker' in drugs trials in future – as we know that it's affected in all forms of FTD and can track the change in volume over time to see if a particular drug is able to slow down the damage to that important part of the brain.

International Research

1. Group Communication Treatment for Individuals with PPA and Their Partners

Lead authors: Aimee Mooney and Melanie Fried-Oken

Where from: Portland State University, Oregon, USA

Where published: Seminars in Speech and Language

When: June 2018

Summary: These researchers from the USA developed a group treatment approach to PPA based on introducing strategies to help people with PPA compensate for their communication difficulties. They used this approach in a group session for five people with different kinds of PPA, as well as their communication partners. The group intervention lasted for six weeks, and treatment sessions were held twice weekly for one hour. Results suggested that the therapy was effective – participants reported gains in knowledge about PPA, and in using different ways to communicate. Moreover, the participants reported feeling more confident about approaching conversations and participating in daily activities. The results suggest that group communication strategies could help people with PPA.

2. Retraining speech production and fluency in non-fluent/agrammatic primary progressive aphasia

Lead authors: Maya Henry and Maria Luisa Gorno-Tempini

Where from: University of Texas and University of California San Francisco, USA

Where published: Brain

When: June 2018

Summary: The authors here looked at the efficacy of video-assisted speech training for people with nonfluent variant PPA – a strategy involving a) structured rehearsals of individualised scripts with a speech and language therapist, and b) intensive home practice using videos. The effectiveness was assessed at 3, 6 and 12 month follow-up visits and the researchers found an overall improvement in production of correct, intelligible scripted words, and a reduction in grammatical errors. Importantly, intelligibility improved both for scripts that the people with PPA had practised, and for new words and phrases as well. The researchers then went on to find out which parts of the brain were supporting this improvement and found that an area in the left middle/inferior

temporal lobe was driving this effect. This area is relatively unaffected in nvPPA and together, the findings here suggest that behavioural interventions can have significant and lasting improvements in speech production in nvPPA.

3. Prevalence of Mathematical and Visuospatial Learning Disabilities in Patients with Posterior Cortical Atrophy

Lead authors: Zachary Miller and Maria Luisa Gorno-Tempini

Where from: University of California San Francisco, USA

Where published: JAMA Neurology

When: June 2018

Summary: These researchers from America reviewed the medical records of 279 people living with dementia in America. Eighty-four had logopenic variant PPA, 100 had typical memory-led Alzheimer's disease, and 95 had a rare visual-led dementia known as posterior cortical atrophy (PCA). The researchers have previously reported that people with lvPPA are more likely to have reported language-based learning difficulties like dyslexia than other people. Here, they added to that finding by showing that people with PCA were more likely to have had a kind of learning difficulty affecting mathematics or visual processing – both of which use the same brain regions that we know are affected in PCA.

These results should be treated with some caution as there is always a danger with retrospective research of a kind of 'ascertainment bias' – and the researchers acknowledge this in their paper. However, if these findings are correct, then they suggest that vulnerabilities in a particular brain network that manifest in childhood as learning difficulties may actually drive the type of dementia that somebody develops later on in life: if somebody has a childhood dyslexia who then develops dementia, perhaps they are more likely to develop lvPPA, whereas if somebody has a learning difficulty affecting their ability to process visuospatial information, they might be more likely to develop PCA.

A really important point is that the research doesn't suggest that developmental learning difficulties are associated with an increased risk of dementia – just that the type of learning difficulty might determine the type of dementia. And again, this is very preliminary research that will need to be replicated.

6. Upcoming Meetings

Upcoming meetings

1. PPA Support Group Meeting

Let us bring your attention to the upcoming **PPA Support Group Meeting**, which will take place on **Thursday 6th September 2018**. During the morning session, **Dr Anna Sobel**, Consultant Psychiatrist, will give an overview of the mood and behavioural challenges that can accompany the initial and later stages of PPA. Her presentation will focus on when to seek support, current treatment options, and practical strategies for the maintenance of good mental health for people with PPA and carers. This will be followed by a short Question and Answers session in which members can ask questions of **Dr Sobel**, or of **Professor Jason Warren**, Consultant Neurologist, who will be chairing the meeting.

We are trialling a new format for the afternoon session: members will be invited to choose one of three broad 'themes' - and we will split into small groups around each theme. We anticipate that there may be several small groups for each theme. Each group will have an experienced member of the support group to help facilitate the discussion, and we will ask for a group spokesperson to summarise the key points of your discussion to the wider group.

The three themes are:

- A) Understanding PPA: receiving a diagnosis;
- B) Practical PPA: finance, paperwork, rights, and finding support;
- C) Living with PPA: activities, relationships and friendships.

Date & Time: Thursday 6th September 2018, 11:00-14:00, arrival from 10:30.

Location: 'The Hub', 5th Floor, Wellcome Collection, 183 Euston Road, London NW1 2BE

To register your place at this event, we kindly ask you to RSVP to Chris Hardy at c.hardy.12@ucl.ac.uk or to leave a message on 07388 220 355. This will help us with guest registration and catering provisions for the light sandwich lunch. In your RSVP, please give the following information:

- 1) If anybody will be accompanying you;
- 2) Which of the three themes you think you would like to discuss during the afternoon session (you won't be tied to this - it's just to give us an idea of numbers);
- 3) Anything we should know (e.g. if this will be your first meeting);
- 4) Any dietary requirements.

2. Joint Carers' Meeting - Monday 5th November 2018, 11:00 - 14:00 (arrival from 10:30). 'The Hub', 5th Floor, Wellcome Collection, 183 Euston Road, London NW1 2BE. RSVP to laura.king@ucl.ac.uk.

3. Bereaved Carers' Meeting - Thursday 15th November 2018, 10:30 - 12:30. 'The Hub', 5th Floor, Wellcome Collection, 183 Euston Road, London NW1 2BE. RSVP to laura.king@ucl.ac.uk.

4. FTD Support Group Meeting - Monday 26th November 2018, 11:00 - 14:00 (arrival from 10:30). 'The Hub', 5th Floor, Wellcome Collection, 183 Euston Road, London NW1 2BE. RSVP to laura.king@ucl.ac.uk.

5. PPA Support Group Meeting - Tuesday 8th January 2019, 11:00 - 14:00 (arrival from 10:30). 'The Hub', 5th Floor, Wellcome Collection, 183 Euston Road, London NW1 2BE. RSVP to laura.king@ucl.ac.uk.

Creating 'stages' of PPA

We also wanted to let you know about some work that we'll be undertaking over the next few months - and something that we'd like your help with. When given a diagnosis of PPA, many people naturally ask what will happen next, how long will they be able to continue with a particular activity, or when and what care will they need. Such questions are often met with the response 'we don't know' or 'it's different for each person'.

In many ways these responses are reasonable, as individual experiences can indeed vary greatly from one person to the next. Symptoms may vary from individual to individual owing to a variety of factors including age, health and the disease underlying the PPA. Not everyone will experience the same symptoms or progress at the same rate. But whilst true, these responses are not terribly helpful.

We would like to create documents for each of the major PPA subtypes that describe the 'stages' that people go through with one of these diagnoses. We hope that the final document that we generate together will be useful for people being diagnosed with PPA in the future.

We have only just been given ethical approval to conduct this research and so there is nothing to do just yet - but we just wanted to take the opportunity to mention it now as it is something that we will write to you about separately in due course.

Rare Dementia Support is a fund held by the National Brain Appeal, the charity dedicated to supporting the work of The National Hospital for Neurology and Neurosurgery and the Institute of Neurology in Queen Square, London. The fund raises money to provide specialist support group services for individuals living with or affected by one of five less common dementia diagnoses.

The services provided by Rare Dementia Support rely on funding from donations, grants and trusts, and supporters taking part in fundraising. The fund aims to raise £150,000 annually in order to maintain and extend the existing support service, and facilitate further access to online resources and information. The ultimate aim is that everyone affected by a form of rare dementia will have access to specialist information and support, as well as contact with other people with a similar condition.

To make a donation to the Rare Dementia Support fund you can [visit our page on JustGiving](#), text RDSF84 plus the amount you would like to donate (e.g. £5) to 70070 or please contact The National Brain Appeal on 020 3448 4724 for details about how to donate by bank transfer.

If you have any queries or ideas or would simply like to know more about the fund for Rare Dementia Support please contact the Fundraising Manager Eva Tait on eva.tait@nhs.net

Thanks to our recent fundraisers!

We want to say a special 'thank you' to Georgie Church, who completed the London Marathon in April this year, raising over £7,000 for Rare Dementia Support. A truly herculean effort in the heat on the day, and we are so grateful to Georgie and to everyone else who has fundraised for us in the past.

Travel costs

We are aware that some people travel long distances to attend our meetings, and that perhaps this isn't always viable from a financial point of view. We have had a small donation from the National Brain Appeal to help with travel costs for meeting attendance for those who need it. Please let Chris know if a contribution towards travel expenses would help to make it possible for someone to attend the meeting.

The term primary progressive aphasia (PPA) refers to a group of degenerative brain disorders (dementias) in which loss of speech and language abilities is the leading and most prominent problem.

'Aphasia' refers to a neurological language problem; 'progressive', becoming worse over time; and 'primary', due to brain tissue changes rather than an external cause.

PPA affects both sexes and usually starts between the ages of 50 and 70. However, it can also affect older people, and rarely, younger people as well.

Within the umbrella term of PPA, three main patterns of language loss are recognised: a nonfluent variant (nfvPPA), semantic variant PPA (svPPA), and logopenic variant PPA (lvPPA).

The nonfluent and semantic variants fall within a larger group of brain disorders collectively called frontotemporal dementia (FTD), indicating the parts of the brain mainly affected: the frontal and temporal lobes. The logopenic variant, by contrast, is usually caused by the same proteins that cause Alzheimer's disease (it is an unusual form of Alzheimer's disease). We also run a frontotemporal dementia support group for people with behavioural variant FTD and all forms of PPA.

In most cases, we do not know what causes PPA in a particular person – in only a small minority of people is the disease caused by an abnormal gene (in contrast to the situation in behavioural variant FTD).

Disclaimer

Please note that you assume full responsibility and risk in the use of information contained on our website, in our newsletters, at support group meetings and in subsequent correspondence. Our support group based correspondence is generic in nature and we are limited in our ability to offer specific advice. We aim to ensure that all information is as accurate as possible but we accept no responsibility for any errors, omissions or inaccuracies, or for any adverse consequences of any kind arising from the use of support group based content. Our regional group facilitators are volunteers. They may refer to regional facilitator guidelines from RDS but are not governed by RDS. They operate independently and with best intention and you assume full responsibility for your contact and engagement with them and in the regional groups they facilitate. Please see the clinician responsible for your care, a social services representative, or your GP if you have specific needs which require attention. Any medical decisions should be taken in discussion with an appropriate healthcare professional.