



Dear PPA Support Group Members,

March 2018

Welcome to the March 2018 edition of the PPA Support Group newsletter. We hope that the recent cold weather hasn't affected you too much, and are looking forward to the arrival of spring as much as we are! In this edition, you'll find information about our next PPA Support Group meeting, to be held on **11th April 2018**, as well as some of the **research developments** coming out of UCL and around the world, and an update from the **National Brain Appeal**.

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 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.

Upcoming Support Group Meeting

Firstly, we wanted to let you know the details of the upcoming **PPA Support Group Meeting**, which will take place on **Wednesday 11th April 2018**. At this meeting, we will hear from neurologist and scientist Dr Thomas Cope from the University of Cambridge, who will give us a talk on some of his recent research looking at why people with "non-fluent" forms of PPA have hearing and grammar problems. He will be joined by Anna Volkmer, who is a highly specialised speech and language therapist who focuses on PPA. Anna has kindly agreed to give us an update on her PhD project, in which she is developing an online therapy resource to support speech and language therapists to deliver communication skills training to people with PPA and their families. The full agenda is on page 2, and we hope to see you there.

Date & Time: Wednesday 11th April 2018, 11:00-14:00 (arrival from 10:30)

Location: 6th floor, Wellcome Trust, Gibbs Building, 215 Euston Road, London NW1 2BE. (As you come through the main entrance of the Wellcome Collection, head up the initial set of stairs. Just past the visitors' reception desk you will see some lifts on the left. Take the lift to the 5th floor and we will then guide you to the correct room). ****Please note the different location to our previous few meetings****

Directions: <https://wellcomecollection.org/visit-us/getting-here>

RSVP: To register your place, we kindly ask you to RSVP to Laura King l.king@wellcome.ac.uk **by Friday 6th April 2018**. This will help us with guest registration and catering provisions for the light sandwich lunch. Please also let us know if you have any special dietary requirements.

AGENDA: PPA Support Group Meeting 11th April 2018

10:30 – 11:00 **REGISTRATION WITH TEA & COFFEE**

11:00 – 11:10 **Welcome and Special Notices**

Dr Chris Hardy, UCL

11:10 – 12:00 **Why do patients with non-fluent PPA have hearing and grammar problems?**

Dr Thomas Cope, University of Cambridge

Dr Thomas Cope is a Clinical Research Fellow and Neurology Registrar at the University of Cambridge and Addenbrooke's Hospital. He conducts research with patients who have neurodegenerative disease, especially those with primary progressive aphasia. He is interested in understanding how the healthy brain 'sees' and 'hears' the outside world, and in explaining how this can go wrong. He explores these questions by combining psychological experiments with advanced brain scanning techniques, especially magnetoencephalography and MRI.

12:00 – 13:00 **LUNCH**

13:00 – 14:00 **Better Conversations with PPA- update on a web based therapy resource for speech and language therapists**

Anna Volkmer, University College London

Anna Volkmer is a highly specialised speech and language therapist who has worked across the UK and Australia. Anna has always worked with adults with communication difficulties and became particularly interested in working with people with primary progressive aphasia (PPA) around 10 years ago. She noticed that there were not many speech and language therapy services who were seeing people with this diagnosis. This motivated her to write a book for other speech and language therapists on how to work with people with PPA. To develop the research in the area of speech and language interventions Anna applied for funding from the National Institute for Health Research to do a PhD. Anna is now in the 3rd year of her PhD developing and piloting an online therapy resource to support speech and language therapists to deliver communication skills training to people with PPA and their families.

14:00 **Formal close of Support Group**

National Brain Appeal Update

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 (<https://www.justgiving.com/campaigns/charity/tnba/rare-dementia-support>), 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

Research Update

We received some helpful feedback from people at our last Support Group meeting, who told us that they felt vaguely aware of the ground-breaking research going on at UCL and around the world, but that they wished they could have a more regular breakdown of research into PPA. To that end, Dr Chris Hardy has kindly offered to distil three pieces of research from UCL and three from around the world for members in our Support Group newsletters. We hope that you find these useful. Chris says:

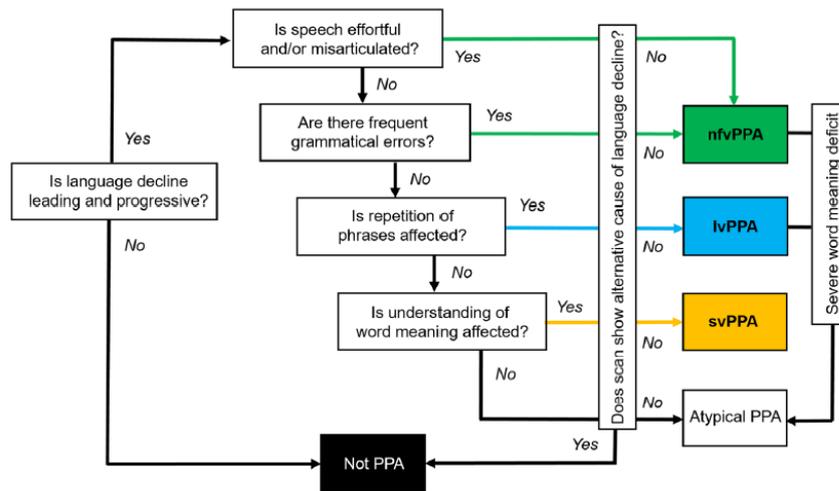
"I promise that I will try to pick papers that I think will be most interesting and relevant to our members – but I may not always get this right – so please do give feedback if I'm straying too far from what would be useful here. The idea is that although we have all of this incredible research going 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!"

UCL

We are conscious that several of you will have been directly or indirectly involved with our research at UCL. Thank you so much – this work would not have been possible without you.

1. **Title:** Primary progressive aphasia: a clinical approach
Authors: Charles Marshall, Chris Hardy, Anna Volkmer, Lucy Russell, Rebecca Bond, Peter Fletcher, Camilla Clark, Jonathan Schott, Martin Rossor, Nick Fox, Sebastian Crutch, Jonathan Rohrer, Jason Warren.
Where published: Journal of Neurology
When: February 2018

Summary: This is a review article, which means that it doesn't report on the results of new experiments or research, but summarises everything that is known about a particular area of research: in this case, primary progressive aphasia. The work here was led by Charles Marshall and Jason Warren, and had input from several other neurologists, psychologists and speech and language therapists. The article is directed at non-specialist clinicians: PPA is very rare (affecting around 3 in every 100,000 people) and this means that many GPs and even some neurologists may not have had much experience with these conditions. Dr Marshall and colleagues attempt to distil all of their years of clinical experience into this article, providing a 'framework' for assessing language that should be really helpful to non-expert clinicians who want to learn more about PPA (included below).



2. **Title:** Plasma tau is increased in frontotemporal dementia

Authors: Martha Foiani, Ione Woollacott, Carolin Heller, Martina Bocchetta, Amanda Heselgrave, Katrina Dick, Lucy Russell, Charles Marshall, Simon Mead, Jonathan Schott, Nick Fox, Jason Warren, Henrik Zetterberg, Jonathan Rohrer.

Where published: Journal of Neurology, Neurosurgery and Psychiatry

When: February 2018

Summary: In this study, which was led by Martha Foiani and Jonathan Rohrer, the researchers took blood samples from people with different kinds of dementia, including behavioural variant frontotemporal dementia, and all three major subtypes of PPA (155 different people in total). We know that different proteins can cause different kinds of dementia, and here the researchers were measuring levels of one of these proteins – *tau* – in the blood samples they took from the participants. Results from the study showed that levels of *tau* in the blood samples were higher in the groups of people with PPA or FTD than in 'control' participants who didn't have any kind of dementia. The authors say that more work is needed to investigate how these *tau* levels change over time, and to see if they might help predict which protein is responsible for causing disease in an individual person. Two people with progressive nonfluent aphasia, for instance, might have very similar symptoms, but these could be caused by problems with completely different proteins. Being able to predict which protein problem an individual person has could be vitally important in knowing which treatment has the best chance of working, as and when treatments do become available.

3. **Title:** Motor signatures of emotional reactivity in frontotemporal dementia

Authors: Charles Marshall, Chris Hardy, Lucy Russell, Camilla Clark, Rebecca Bond, Katrina Dick, Emilie Brotherhood, Cath Mummery, Jonathan Schott, Jonathan Rohrer, James Kilner, Jason Warren.

Where published: Scientific Reports

When: January 2018

Summary: When we see somebody else smiling or laughing, we will unconsciously mimic those same emotions – a phenomenon known as "automatic motor mimicry". We know that people with different kinds of frontotemporal dementia (including some of the PPAs) have problems with understanding emotions, and Charles Marshall and Jason Warren, who led this research, wanted to find out if this automatic motor

mimicry might be going wrong in different kinds of FTD, and if this might explain why some people find it harder to understand emotions after diagnosis. The researchers studied facial muscle responses of 37 people with different kinds of FTD and 21 people without dementia as they watched videos of emotional facial expressions. People without dementia showed automatic imitation of these emotions, but this wasn't seen in people with the behavioural variant of FTD. People with semantic dementia did show these automatic responses, but did not show the normal 'coupling' between motor responses and correct emotion identification (e.g. "That person is happy"). This is the first time that this has been investigated in FTD, and although further research is needed, the authors are hopeful that this kind of approach might help with diagnosis, and could potentially pave the way for retraining of emotional responsiveness in people with different kinds of FTD.

Around the World

- Title:** [18F]AV-1451 tau-PET and primary progressive aphasia
Authors: Josephs KA, Martin PR, Botha H, Schwarz CG, Duffy JR, Clark HM, Machulda MM, Graff-Radford J, Weigand SD, Senjem ML, Utianski RL, Drubach DA, Boeve BF, Jones DT, Knopman DS, Petersen RC, Jack CR Jr, Lowe VJ, Whitwell JL.
Where from: Mayo clinic, USA
Where published: Annals of Neurology
When: February 2018
Summary: 'PET' or positron emission tomography is a special kind of brain scan in which a 'tracer' is injected into the body and binds to a particular protein in the brain. Here, the authors used a special kind of tracer that binds to *tau*, and they were able to look at the brains of people with all different kinds of PPA to see if and where in the brain they could see elevated levels of *tau*. Across 40 people with PPA, they showed that different patterns were observed depending on the clinical subtype: in logopenic aphasia, there was increased binding in the temporal and parietal lobes; in semantic PPA there was increased binding in the left temporal lobe, further forward than seen in logopenic aphasia, and in progressive nonfluent aphasia, there was higher binding to *tau* in regions right at the front of the brain and also within deeper 'subcortical' structures. All in all, this particular tracer was excellent at distinguishing between the PPA variants, and the authors argue that it could be useful in helping to diagnose somebody accurately in future.
- Title:** Neuropsychological differentiation of progressive aphasic disorders
Authors: Harris JM, Saxon JA, Jones M, Snowden JS, Thompson JC.
Where from: University of Manchester, UK
Where published: Journal of Neuropsychology
When: February 2018
Summary: It can be really difficult for clinicians to accurately diagnose PPA, and identifying which specific subtype somebody has is particularly challenging. People are often asked to have neuropsychological testing to help clinicians to make a diagnosis, and the researchers here wanted to find out which specific tests are most useful in discriminating between syndromes. They studied 47 patients with different kinds of PPA and typical Alzheimer's disease, and found that tasks assessing sentence ordering, producing speech, and making coordinated movements with the face were particularly useful in identifying progressive nonfluent aphasia. A test of understanding single words was most useful in detecting semantic dementia, but no tests were particularly useful in identifying logopenic aphasia, which the authors say emphasizes the difficulties in classifying this subtype.
- Title:** Rates of amyloid imaging positivity in patients with primary progressive aphasia
Authors: Santos-Santos MA, Rabinovici GD, Iaccarino L, Ayakta N, Tammewar G, Lobach I, Henry ML, Hubbarly I, Mandelli ML, Spinelli E, Miller ZA, Pressman PS, O'Neil JP, Ghosh P, Lazaris A, Meyer M, Watson C, Yoon SJ, Rosen HJ, Grinberg L, Seeley WW, Miller BL, Jagust WJ, Gorno-Tempini ML.

Where from: University of California San Francisco, USA

Where published: JAMA Neurology

When: January 2018

Summary: The researchers in this study used PET imaging (as in the study above), but here they used a different tracer known to bind to a protein called *amyloid*. We know that this protein is involved early on in some kinds of dementia, including Alzheimer's disease. The authors wanted to understand more about *amyloid* in PPA and so used this tracer in 89 people with PPA. Twenty-four out of 28 people with semantic dementia had scans that were negative for *amyloid*; 28 out of 31 with progressive nonfluent aphasia had negative *amyloid* scans, but 25 out of 26 people with logopenic aphasia had positive *amyloid* scan results. Importantly, several of the people who participated in the research had donated their brains after they died. This meant that the scientists could look directly at the brains of the people who had slightly different *amyloid* results to the other people in their syndromic group. The results confirm previous work suggesting that *amyloid* PET scanning could be useful in diagnosing LPA.

Contact us!

We look forward to bringing you more of what we hope is useful and interesting PPA-specific information in future newsletters; please do let us know if you have anything that you would like to share. We remain completely committed to supporting people with a diagnosis of PPA wherever possible, and hope to see as many of you as can make it at our meeting on April 11th. If you need or want to get in touch before then, please do so and we will be glad to help.

With Best Wishes

The PPA Support Group Team

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 via this means. 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 health care professional.

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