

PCA SUPPORT GROUP

Newsletter

Welcome to the PCA Support Group Newsletter

Issue 4 January 2009

A belated Happy New Year from all here at the National Hospital. We hope this newsletter finds you looking forward to the year ahead, and that we will see as many of you as are able and keen to attend at the three PCA Support Group meetings scheduled for 2009 (13th March, 10th July, 12th November).

PCA Support Group Meeting 21 November 2008

At the last group meeting in November, we enjoyed a presentation from Clare Taylor of the Alzheimer's Society. Clare is the Living with Dementia Coordinator. She works directly with people with a diagnosis of dementia, in whatever form it takes. The key aim of her role is to ensure that all people with dementia have a voice within the Alzheimer's Society and in wider society. She also works to champion the rights of people with dementia and to influence the policies and services that affect them.

Clare explained that she runs the 'Living with Dementia' programme, to which a number of people with dementia and their family members volunteer. Volunteering can take multiple forms:

- Giving presentations and raising public awareness
- Contributing articles and letters to the Living with Dementia monthly magazine
- Lobbying MPs and commenting on government legislation
- Being interviewed by national press and television
- Recruiting and inducting new staff at the Alzheimer's Society
- Helping to make the website easier to use
- Developing information for other people with dementia and their families
- Participating in the national consultative body, the Living with Dementia Working group

There is potentially a huge range of local and national activity that people can engage in. If you would like more information about the scheme, please contact Clare Taylor on 020 7423 3591 or at clare.taylor@alzheimers.org.uk

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PCA received some further welcome attention and coverage in the most recent edition of the Alzheimer's Society magazine *Living with Dementia*. In the article, fantasy author Terry Pratchett and our very own Graham Doggett discuss some of their experiences of and reactions to their own PCA. In case you missed it, a copy of this article is attached to the end of this newsletter. Terry Pratchett also wrote another article for a professional journal, *Advances in Clinical Neuroscience and Rehabilitation*. This is also attached below for your interest.

PhD Studentship on PCA

We are delighted to announce that the Dementia Research Centre has won funding from the Alzheimer's Society for a PhD Studentship project aimed at better understanding the brain changes and visual problems experienced in PCA. The award to Manja Lehmann, who many of you will know from our PCA Support Group meetings, will be supervised by

Nick Fox, Seb Crutch and Jo Barnes.

This represents one of the very first research projects dedicated to PCA to be funded in the UK.

PCA factsheet

Thank you very much for all your feedback on the factsheet 'What is posterior cortical atrophy'. Your suggested improvements have been added, and the sheet is due to be published in print and online by the Alzheimer's Society in early 2009.

On-line blogs

Fear not, if the term on-line blog means nothing to you! Graham Doggett has been very busy creating an internet-based record of some of his thoughts and experiences regarding PCA. The blog currently offers some diary excerpts and copy of an article about Graham and his wife Susan which appeared in *The Daily Telegraph* in December 2008.

As Graham says on the blog: "I wanted to

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create this blog to share strategies that may be useful for me and others similarly afflicted for coping with the bizarre effects of PCA - for example ways to cope with using telephones, computers, diaries, as well as reading and answering printed letters/mail, shopping, public transport, dressing oneself etc and generally getting through life in a world that is not usually

accommodating for someone who has visual problems but is not blind in the conventional way.”

You can add your own comments and reflections to the blog. Graham's blog can be found at:

<http://pcachat.blogspot.com/>

Next PCA Meeting:

Friday 13 March 2009

Royal London Homeopathic Hospital Conference Suite,
60 Great Ormond Street, London, WC1N 3HR.

At the March meeting:

- **Riitta Kukkastenvemas** talking about financial benefits available to those with PCA
- Question and Answer session with **Prof Nick Fox** and **Dr Cath Mummery**

Plus, future meeting dates for your diaries:

Friday 10 July and Friday 13 November 2009

Contact Us:

Jane Douglas 08451 555 000 x 723560 or email jdouglas@drc.ion.ucl.ac.uk

Sebastian Crutch 08451 555 000 x 723113 or email s.crutch@drc.ion.ucl.ac.uk

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Sharing the experience of PCA

In the latest in our series about sharing personal experiences of PCA, one young carer whose mother has PCA describes some of their story, and tells of ideas and strategies they have found useful in coping with the condition.

Mum was placed under a Private General Geriatrician for two years, but they did not pick up on Mum's reading and writing going so early. The geriatrician did however get Mum onto Aricept very quickly - just in time to be covered by the NHS.

After the two years, a friend of mine suggested we see Professor Fox. The tests performed by Professor Fox and his team really homed in on Mum's difficulties and these tests should be more widely used to identify that people do need to be referred to a Neurologist. I do not believe that PCA or Alzheimer's can be treated successfully while within private health

care. It is much better to be treated within the NHS as this opens up lines of local support and benefits.

A few practical devices and ideas that we have found useful:

- I have had some **handbands** made with a website address and our road and postcode on them. The website gives alternate emergency contact details if I am involved in an accident. The road and postcode allows Mum to get taxis if she gets lost. She is very afraid of getting lost and not being able to tell someone where she lives. I wanted to avoid having full details in her purse, but she would get old addressed envelopes and have those in her purse instead. In the end I created a laminated note with her full details and she now has that in her purse.
- **Meditags** are useful. Both Mum and I wear one. Mine refers to a website with alternative emergency contact details. Mum is registered with Meditag.
- Her **taxicard** is very useful and cost effective. But, always give a note to the person as to where they are going. The taxi drivers want to confirm they have the right passenger and sometimes may not have the full details of the destination.

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- **Bagging up** medication on a regular basis has helped keep tabs on what mum has taken. I 'bag up' every 28 days and put out 7 days medication at a time. This includes all the medical medication as well as the vitamins and supplements. I use small Ziploc bags that one can get at Ryman's or Staples and clearly label them as 'Monday Breakfast' or 'Monday Supper' etc.
- Doro **Careline Telephone** plus keypad extension key pad very good (bought via RNIB website).
- Simple, daily written/printed reminders worth having to use each day. Large readable fonts. Mum often is concerned about what is happening and when.
- The **speaking clock** I bought from the RNIB site has really helped. Two presses gives the day of the week and date.

People

- Try, as much as possible, to involve close family and friends. Mum helps a friend in a charity shop on a regular basis and enjoys the company. Luckily my mother's brother looks after Mum if I need to go away for a weekend.
- A volunteer visitor is helping Mum read a newspaper on a weekly basis.
- Group holidays are good, but let others know that the person you

are with may need help with certain things. They are normally happy to help where they can.

- If someone is taking Mum out, I need to ask them to ensure that Mum is helped down stairs and guided to the rails.

Food and shopping

- Mum likes to go to the local fish & chip shop to get hot food. She cannot use any cooking appliances in the kitchen, the kettle is a challenge, but most of the time she manages to get some hot coffee! I have considered Meals on Wheels – but Mum is fairly mobile still and enjoys her freedom. We eat our main meal in the evening, which I cook – I am getting better! Luckily Mum likes cold meat and tomatoes and I am now trying to keep the fridge less crowded so Mum can see what is available for lunch. Due to reading difficulties, she is a bit worried about the dates on foods?
- Mum is on a vitamin and supplement regime based on suggestions from the Brain Bio Centre. I am not sure if they make a difference. We have stopped taking certain supplements for a short time and maybe mum deteriorated slightly – this may be my mind playing tricks? I believe that the supplements do supplement her diet – her appetite

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for fruit and vegetables is not great and hence, I believe, multi-vitamins at the least are worthwhile.

Other things to consider

- Mum is afraid of the dark now – one needs to factor that in in any plans or journeys.
- PCA tends to be a young person's disease, whereas AD mostly affects older people. Many of the groups that Mum has tried have comprised people much older than herself, making her feel less comfortable.

Dementia the 'jokester'

Fantasy author Terry Pratchett and former quantum chemist Graham Doggett both have posterior cortical atrophy (PCA). Here are extracts from their accounts of life with a rare form of Alzheimer's

Since being diagnosed with the condition just over a year ago, Terry Pratchett has often emphasised the need to talk about dementia in order to tackle the stigma. At the end of the third annual UK Dementia Congress held in Bournemouth in October, the author gave a speech about having PCA.

Speaking about his rare diagnosis, Pratchett says, 'If I didn't tell you I had it you wouldn't know. That is the signature of PCA; for a while at least it leaves you quite free to talk. The key problem for me, and the one that first led me to go to the doctor more than a year ago, was that my typing and spelling were both becoming quite atrocious. This is not good for a professional author who has touch typed since he was 13.

'The simple way to describe what happens is this: it's like working in a room where a light flickers on and off. Sometimes, when it's on, I can spell long words like transubstantial and marmalade. When it is momentarily off I sometimes lose track of how to spell a word as simple as elf.

'There are other, more standard problems; one feels like one is followed around all the time by an invisible moron who loses things and does things wrong, when you have thought you were doing them right. You end up not trusting yourself, although I have to say, things do become a little easier when you learn a few tricks.

'PCA laughs at you; it's a jokester. I was speaking to a man with PCA who conversed in the tones and

vocabulary of a university lecturer but could not recognise his teacup on the table. At the same meeting, I met a lady who was beautifully assured in her speech but cannot dress herself. Her brain cannot handle the topological problems involved.

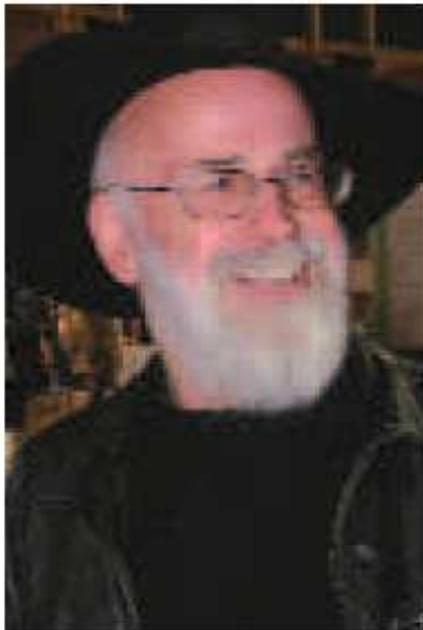
'I am experiencing this also; in a small way as yet. Again this is typical PCA. The eyes may be working fine. The brain, however, has difficulty with the labelling.'

On the subject of his diagnosis, Pratchett says, 'If you are given a diagnosis with the word Alzheimer's in it, the word immediately glows bright red and you barely hear anything else the specialist says. But when I had got to grips with the idea of PCA, there was no specialist in my area prepared to take me on. Even among healthcare professionals PCA seems to be something of a mystery.'

Graham Doggett

I have PCA, a rare form of Alzheimer's which affects the part of the brain where visual messages are decoded. I am registered blind.

 Intellectually my brain still functioned at a good level but my visual problems were debilitating. I was forced to retire from a job I loved. 



Terry Pratchett, left, pictured at the UK Dementia Congress (picture courtesy of Geoff Hodgson) and Graham Doggett

I am unable to read; use a computer (after doing so for 50 years); read my own writing; use the keys on a phone; watch TV or films; recognise people (including my wife of 47 years at times). I cannot read body language or judge the speed of vehicles when crossing the road. Shopping, dressing, gardening; anything requiring binocular vision is challenging.

My problem first became apparent in 2003 when I had difficulty reading students' work. At the time I was working on two quantum chemistry projects with colleagues overseas. I was distraught to realise that I could not continue with the research. Intellectually my brain still functioned at a good level but my visual problems were debilitating.

At the age of 63, I was forced to retire from a job I loved. An optician's field test revealed problems and I was sent to an ophthalmologist. He was puzzled by my symptoms and unable to make a diagnosis. On moving to Hertfordshire, a different ophthalmologist sent me for a

head scan which revealed a neurological problem.

I was subsequently advised I had Alzheimer's disease. For over eighteen months we were left in limbo. Eventually a relative in America emailed us a paper entitled Benson's Syndrome which described my curious symptoms and mentioned the name posterior cortical atrophy.

At last I felt we were on the right tracks but still I was having no treatment. Our daughter suggested I ask to be referred to the National Hospital for Neurology in London. When we saw the consultant she diagnosed PCA and prescribed Aricept.

When people are told I have Alzheimer's they find it difficult to understand how I can converse fluently on many topics. They realise my problems when I attempt to open a door and climb into their car! Incidentally I don't like being labelled as someone with dementia. My dictionary definition states, 'species of insanity consisting in feebleness of mind.'

Quick facts

What is PCA?

Posterior cortical atrophy (PCA) is a progressive degenerative condition involving the loss and dysfunction of brain cells, particularly at the back (posterior) of the brain.

In the vast majority of cases, this loss of brain cells is associated with the same pathological brain changes seen in typical Alzheimer's disease, so PCA is usually considered to be an atypical variant of Alzheimer's.

PCA and Alzheimer's disease affect people in very different ways. Alzheimer's disease is most commonly associated with deterioration in memory, language and perception. By contrast, people with PCA tend to have relatively well preserved memory and language but instead experience a considerable decline in visual abilities, for example identifying and locating objects and in skills such as spelling, writing and arithmetic.

The Society is producing a factsheet on PCA which will be available next year.

Living with Dementia



Terry Pratchett, OBE

Terry Pratchett is one of the most popular authors writing today and is the acclaimed creator of the bestselling Discworld series. The first Discworld novel, *The Colour of Magic*, was published in 1985 and there are now 36 books in the series. The first Discworld novel for children, *The Amazing Maurice and his Educated Rodents*, was published in 2001 and was awarded the 2001 Carnegie Medal.

Long regarded as a significant satirist, Pratchett has won numerous literary awards, was appointed OBE in 1998 and has received four honorary doctorates.

Terry Pratchett Featured in New Year Honours

Author Terry Pratchett was awarded a knighthood for services to literature in the New Year Honours list. The best-selling fantasy writer has sold almost 60 million books worldwide.

Neil Hunt, Chief Executive of Alzheimer's Society, says: "I would like to congratulate Terry on this fantastic and well-deserved achievement. Terry is not only a successful author and well-loved public figure, he is also playing a key role in fighting the misunderstanding and stigma surrounding dementia."

Seven hundred thousand people who have dementia in this country are not heard. I'm fortunate, I can be heard. Regrettably, it's amazing how people listen if you stand up in public and give away \$1 million. This report goes some way to redressing that situation and allows others to tell you all about dementia. I regarded finding I had a form of Alzheimer's as an insult and I decided to do my best to marshal any kind of forces that I could against this wretched disease.

I have posterior cortical atrophy or PCA. They say, rather ingenuously, that if you have Alzheimer's it's the best form of Alzheimer's to have. This is a moot point, but what it does do, while gradually robbing you of memory, visual acuity and other things you didn't know you had until you miss them, is leave you more or less as fluent and coherent as you have always been.

I spoke to a fellow sufferer recently (or as I prefer to say 'a person who is thoroughly annoyed with the fact they have dementia') who talked in the tones of a university lecturer and in every respect was quite capable of taking part in an animated conversation. Nevertheless, he could not see the teacup in front of him. His eyes knew that the cup was there; his brain was not passing along the information. This disease slips you away a little bit at a time and lets you watch it happen.

When I look back now, I suspect there may be some truth in the speculation that dementia (of which Alzheimer's is the most common form) may be present in the body for quite some time before it is capable of diagnosis. For me, things came to a head in the late summer of 2007. My typing had been getting progressively worse and my spelling had become erratic. I grew to recognise what I came to call Clapham Junction days when the demands of the office just grew too much to deal with. I was initially diagnosed not with Alzheimer's but with an ischaemic change, a simple loss of brain cells due to normal ageing. That satisfied me until the next Clapham Junction day. I went back to my GP and said that I knew there was something more going on.

Fortunately, she knew well enough not to bother with the frankly pathetic MMSE test and sent me to Addenbrookes Hospital, Cambridge when, after examination of my MRI scan and an afternoon of complex testing, I was diagnosed with PCA – an uncommon variant which had escaped the eagle eyes of the original diagnostician.

When Milton's Satan stood in the pit of hell and ragged at heaven, he was merely a trifle miffed compared to how I felt on that day. I felt totally alone with the world receding away from me in every direction and you could have used my anger to weld steel. Only my family and the fact I had fans in the medical profession, who were

able to give me useful advice got me through that moment. I feel very sorry for, and angry on behalf of, the people who don't have the easy ride I had. It is astonishing how long it takes some people to get diagnosed (they write to me). I cannot help but wonder if this is because doctors are sometimes reluctant to give the patient the stigma of dementia since there is no cure.

I was extremely fortunate in my GP. I think she was amazed to find that of the two specialists in my area, one had no experience of PCA and therefore did not feel he could not help me and the other would only take on patients over 65 – I was clearly too young to have Alzheimer's. I remember on that day of rage thinking that if I'd been diagnosed with cancer of any kind, at least there would have opened in front of me a trodden path. There would have been specialists, examinations, there would be in short, some machinery in place. I was not in the mood for a response that said, more or less, 'go away and come back in six years'.

My wife said 'thank goodness it isn't a brain tumour' but all I could think then was 'I know three people who have got better after having a brain tumour. I haven't heard of anyone who's got better from Alzheimer's'.

It was my typing and spelling that convinced me that the diagnosis was right. They had gone haywire. Other problems I put down to my looming 60th birthday. I thought no one else had noticed the fumbling with seat belts and the several attempts to get clothing on properly, but my wife and PA were worrying.

We still have the occasional Clapham Junction days, now understood and dealt with. I have written 47 novels in the past 25 years, but now I have to check even quite simple words – they just blank on me, at random. I would not dare to write this without the once despised checker, and you would have your work cut out to read it, believe me.

On the other hand – and this is very typical of PCA – when the kind lady who periodically checks me out asks me to name as many animals as I can, I started with the rock hyrax, the nearest living relative to the elephant, and thylacine – the probably extinct Tasmanian marsupial wolf. That's the gift or the curse of our little variant. We have extreme problems handling the physical world but we can come pretty close to talking our way out of it so you don't notice. We might have our shirts done up wrong, but might be able to convince you it's the new style.

I felt that all I had was a voice, and I should make it heard. It never occurred to me not to use it. I went on the net and told, well, everyone. I wish I could say it was an act of bravery; it wasn't and I find that suggestion very nearly obscene.

How brave is it to say that you have a disease that does not hint of a dissolute youth, riotous living or even terrible eating habits? Anyone can contract dementia; and every day and with a growing momentum, anybody does.

It occurred to me that at one point it was like I had two diseases – one was Alzheimer's and the other was knowing I had Alzheimer's. There were times when I thought I'd have been much happier not knowing, just accepting that I'd lost brain cells and one day they'd probably grow back or whatever. It is better to know, though, and better for it to be known, because it has got people talking, which I rather think was what I had in mind. The \$1million I pledged to the Alzheimer's Research Trust was just to make them talk a bit louder for a while.

It is a strange life, when you 'come out' people get embarrassed, lower their voices, get lost for words. Journalists, on the other hand – I appreciate that other people living with the disease don't get so much of this – find it hard to talk to me about anything else, and it dominates every interview. Yes, I said I had PCA ten months ago, yes, I still have it, yes, I wish I didn't, no, there is no cure. I can't really object to all this, but it's strange that a disease that attracts so much attention, awe, fear and superstition is so underfunded in treatment and research.

We don't know what causes it, and as far as we know the only way to be sure of not developing it is to die young. Regular exercise and sensible eating habits are a good idea but they don't come with any guarantees. There is no cure. So we hope – more hope than would fit in Pandora's box, where it was the last thing.

We hope very carefully that a half-way cure will arrive. Researchers are talking about the possibility of a whole palette of treatments or regimes to help those people with dementia to live active and satisfying lives with the dis-

ease kept in reasonably permanent check in very much the same way as treatments now exist for HIV. Not so much a cure therefore as – we hope – a permanent reprieve. We hope it will come quickly, and be affordable.

In the meantime we hope for Aricept, which is not a cure but acts as a line of sandbags against the rising tide of unknowing. However, it is available free only to those in the moderate stages of the disease: others must pay \$1,000 a year, which I do. Eligibility is determined by the MMSE test, and it would be so easy for a patient in the mild stage to cheat their score into the free zone that I take my hat off to those too proud or responsible to do so. I cough up. NICE says the change it makes at my stage is minimal, but we don't think so in our house, where those little changes make the difference between a dull day and a fine day. The disease is, after all about small changes, and it may be that individuals may indeed be individual.

And that is nearly it for hope at the moment. When my father was in his terminal year, I discussed death with him. I recall very clearly his relief that the cancer that was taking him was at least allowing him 'all his marbles'.

Dementia in its varied forms is not like cancer. Dad saw the cancer in his pancreas as an invader. But Alzheimer's is me, unwinding, losing trust in myself, a butt of my own jokes and on bad days capable of playing hunt the slipper by myself and losing. You can't battle it, you can't be a plucky survivor. It just steals you from yourself.

And I'm 60; that's supposed to be the new 40. The baby boomers are getting older and will stay older for longer – will expect to stay younger for longer. And they will run right into the dementia firing range. How will a society cope? Especially a society that can't so readily rely on those stable family relationships that traditionally provided the back-

bone of care in previous generations?

What is needed is will and determination. The first step is to talk openly about dementia because it's a fact, well enshrined in folklore; that if we are to kill the demon then first we have to say its name. Once we have recognised the demon, without secrecy or shame, we can find its weaknesses. Regrettably, one of the best swords for killing demons like this is made of gold – lots and lots of gold. These days we call it funding.

I believe that the D-day battle on Alzheimer's will be engaged quite shortly and a lot of things I've heard from experts in the field, not always formally, strengthen that belief. It is a physical disease, not some mystic curse; therefore it will fall to a physical cure. There's time to kill the demon before it grows.

I want to thank the Alzheimer's Society for publishing this report and bringing closer the day when the funding we need is made available. This report is part of the Society's Living With Dementia programme, which I'm pleased to see has the support of Comic Relief. A member of the Society once said at a conference: 'I am not dying of dementia – I am living with dementia'. And so, the programme was born; to help those with dementia tell it like it is to the rest of the world and help influence for the better the lives of all of us with this 'embarrassance'. ♦

Terry Pratchett's comments came at the launch of *Dementia - Out of the Shadows*, a new report into the impact stigma has on the lives of people with dementia.

The report and videos featuring four people with dementia sharing their views on stigma and diagnosis can be viewed at www.alzheimers.org.uk/outoftheshadows.



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