111 | Clinical Case 15 – with Dr. Wendy Kelso

January 15, 2023

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Speakers: Wendy Kelso, Ryan Van Patten, John Bellone

Intro Music 00:00

John Bellone 00:17

Welcome, everyone to Navigating Neuropsychology of voyage into the depths of the brain and behavior brought to you by INS. I'm John Bellone.

Ryan Van Patten 00:26

And I'm Ryan Van Patten. And we are board certified neuropsychologists. Today we speak with Dr. Wendy Kelso about a clinical case. This is a woman with posterior cortical atrophy, or PCA. Wendy is a senior clinical neuropsychologist and coordinator of neuropsychology services at the Royal Melbourne Hospital in Victoria, Australia. The first 10 minutes or so of the interview, consist of Wendy's review of the case. And then we begin

Special thanks to Hunter Holoubek & Shanna Cooper for transcribing this episode.

discussing PCA more broadly bringing in additional information from our clinical case when relevant. You'll notice that my audio sounds a bit muffled or distorted during the episode. This was due to a temporary tech issue. I believe that everything is readily understandable, but We apologize for any inconvenience. So with that, we give you our discussion with Dr. Wendy Kelso.

Transition Music 01:19

Wendy Kelso 01:28

This patient is a 60 year old right-handed English-speaking Caucasian female with 18 years education. She was married with three children, and was referred for diagnostic assessment on a background of a three year decline in cognitive functioning and increased anxiety while working as a schoolteacher. She had initially been seen by an ophthalmologist and then by a neurologist, who referred her for further investigation. She attended the interview with her husband and she reported subtle cognitive changes over the past three years, with more marked changes in the past 12 months. Initial symptoms included transient floaters in her eye for which she had sought consultation without the ophthalmologist. And at that time, visual fields were full and the ophthalmological examination was essentially normal. The patient had experienced heightened anxiety when teaching the classroom and in crowded situations. And more recently, she noticed difficulties with completing complex mathematical equations. She'd also noticed subtle changes when trying to recognize the faces of family and friends, and occasionally misperceived on misidentified objects and was unable to recognize some. She was still able to use a mobile phone however, she was finding it increasingly difficult to locate the numbers and the apps that she required.

Over the past year, she'd had greater difficulty with reading and spelling. She had been an avid reader, but was now having difficulty following the line in the text of the book. She would often put a piece of paper or object underneath each line to visually track it while reading, but of note that she didn't have any difficulty recalling information that she'd previously read. For the past six months, she'd had significant difficulties when teaching and these included doing a numerical operations such as misplacing decimal points, difficulties with simple calculations, and also having difficulties when using a computer keyboard where she wasn't able to locate the letters when she was typing. No one had really noticed any significant difficulties with her and the people that she was teaching with had not noticing any cognitive changes. She was having difficulty reading an analog watch, and having more difficulty calculating changes when she was shopping. And a notable change was that she was an avid ballroom dancer, and when she was dancing with other her partner, she wasn't able to put her body in the right position in space to be able to dance well with her partner. She didn't report any changes to memory functioning, and she was able to remember conversations and contents of books and birthdays and other events without difficulty.

When we spoke to her husband, he corroborated the history the patient provided and said the most notable changes were related to mathematical ability, including difficulties with mental arithmetic and calculating decimal places. He'd also noticed the occasional word difficulties. The husband described the patient as the same person with no notable changes in personality or mood. In terms of activities of daily living, she was still

able to do most things but her husband had taken over managing the finances several years ago as he had retired. Her children had not noticed any cognitive changes and believed that the symptoms that she was expressing were related to heightened anxiety, and were wondering if anxiety and excessive worry were the cause of her current problems. She continued to drive how she'd become much more nervous driving and when she was a passenger, and she become much more sensitive to glare on the roads and low light conditions, and was finding it more difficult to judge distance of the cars around her. She'd become disorientated just one time in a shopping center that she wasn't familiar with, where she wasn't able to recognize some of the landmarks that would have previously helped her. And she was also much more nervous using escalators at shopping centers and train stations as she was having difficulty the depth perception and she wasn't sure how far to move her feet up and down when going up and down stairs, and she felt that she might fall into abyss when she was at the top of the escalator.

With regards to psychiatric history, she described herself as a worrier, but had no previous psychiatric history. However, over the last past several years, there was a significant increase in anxiety with symptoms of panic in crowded and unfamiliar environments. She was uncertain to why she was much more anxious. There not been any notable trigger or change in life circumstance.

Of note there was a family history of early onset dementia in an aunt, but no other family history in either parents'. Brain MRI revealed biparietal and occipital atrophy and borderline for age widening of the choroidal fissures. And she had a SPECT scan at the time which revealed mild superior posterior parietal and occipital hypoperfusion bilaterally. And of note her amyloid PET was positive for Alzheimer's disease. In terms of CSF and plasma biomarkers, she had plasma neuro filament light chain, which is an indication of breakdown in axons indicating neurological disease, and that was 52 and the normal range is less than 15. In terms of the CSF biomarkers for Alzheimer's disease, she had a reduction in a-beta and increased phospho-tau and total tau, which was consistent with the presence of Alzheimer's disease.

In terms medication, she was taking cholesterol-lowering medication but otherwise wasn't on any other medications. In terms of the presentation, she presented as a polite and well orientated, middle aged lady who is appropriate in all social interactions. Of note that she found it difficult when she walked into the room to position herself in the chair and arrange herself in the desk.

Premorbid ability was in the higher average range, she was orientated to person, place and time and knowledgeable of reasons and events. Immediate auditory attention span was mildly reduced in the average range and working memory ability was below expected levels in the below average range. Her speed of processing was in keeping with expectations on all the verbally-mediated tasks. But she had great difficulty on psychomotor speed tasks such as the Coding subtest and Symbol Search. And that was mainly because of difficulties with visual tracking and big hour to also shift her gaze up and down. In terms of her performance on memory tasks, her visual and verbal memory ability was at low end of the average range. She performed pretty normally on all verbally mediated tasks, and because of her significant difficulty with visuospatial skills, it was quite hard to get a reliable estimate of visual memory. But I use the Warrington Facial Recognition Memory Test, which is a test where you have a variety of different faces, and then you have to choose which one you saw out of two immediately afterwards. And she actually performed normally on that test indicating preserved memory

function. In terms of visual spatial skills, they were the most significantly affected out of all of the different domains with moderate to severe reductions across most measures. She had very significant spatial difficulties on the Rey Complex Figure where it was very specially distorted, great difficulty on block design, and even doing simple copy, she's able to copy sort of a flower and arrows and some steps. But for example, when copying, some of the flowers had extra petals, she forgot the wheel on one of the bikes. And when she was asked to copy a clock face, she actually put all the numbers outside the clock and labeled the numbers from 1 to 14, which was grossly abnormal for someone that was still working as a teacher at the time. I also gave her some more sensitive tests of visual spatial dysfunction. So the Visual Object and Space Perception Battery, or the VOSP, where she was given object decision and silhouettes where you have to work out which one is is an object and also work out what something is from just an unusual angle such as a silhouette of an object. And she failed both of those tests. She also had difficulty completing a star cancellation test, which took her about four minutes to complete. And she was able to bisect, about four out of the six lines. Prior neuropsychiatric examination revealed finger agnosia, dyscalculia, and dyspraxia. In terms of language functioning basically, his speech was fluent, she had very occasional word finding difficulties, but her performance and all formal language tasks was normal. She had difficulties with reading, so you can read individual words and paragraphs, but she'd sometimes miss words at the beginning or end of a paragraph. And she'd actually have to put a little bit of paper above and below so she could read the line clearly, or track with her finger. In terms of executive functioning, that was all fairly much in the average range, the the main difficulties are really on visual spatial and visual constructional ability. And social cognition was preserved. And her mood showed sort of moderate anxiety, but otherwise she wasn't depressed and didn't have any significant stress going on in their life at that time.

Ryan Van Patten 11:16

Thank you. So we were talking about posterior cortical atrophy here and to my era, the case you described is a really, really good exemplar case of PCA as we think about what PCA entails. So neuropsychologists spend a lot of time thinking about the frontal and temporal lobes of the brain and rightly so. But today, we'll really focus on the parietal and occipital lobes, where PCA tends to hit. So just real quick PCA is in neurodegenerative syndrome, primarily affecting occipital and parietal lobes. It is most commonly caused by Alzheimer's disease pathology, but it's not exclusive to AD. So dementia with Lewy bodies, other Parkinson's plus syndromes, even prion diseases occasionally or rarely can cause PCA. The point here being PCA is not always only AD pathology, multiple pathologies can cause the syndrome. But those pathologies will be posterior in the brain. It's usually early onset in terms of these later syndromes. So maybe between ages 50 and 65, roughly speaking, which fits with your patient. And I just want to highlight a few things that came out to me as you're talking, Wendy. So a lot of the symptoms you described, I think, are very helpful as we start to think about what type of symptoms and functional problems people with PCA might have. So, of course, posteriorly there's going to be visual, visual spatial problems. The patient is not blind, as in their eyes work, but the occipital lobe and all the v1 and visual association areas are really affected. So some of those deficits might be a little more straightforward in our minds, but there's also parietal pathology. So things like the features of the Gerstmann syndrome, Balint's syndrome, we can get into all that that kind of thing comes in more frequently. Apraxia, you mentioned she was a ballroom dancer, and had trouble with the sequenced movements of ballroom dancing. So apraxia come on board, you mentioned that the problems reading, which we think of as a verbal language-based skill, but

obviously requires official spatial skills to read well, and when, when there's so much severe pathology in the back of the brain, even reading can be impaired. So we need to think a complex way about this, right? If somebody scores low on a test of single word reading, they still could have PCA in fact, PCA could be the cause of that. So we don't want to be overly simple with how we think about our tests. I think there was a few of the big topics I just wanted to highlight from what you said. So let me ask a question. What if you step back, generally thinking about PCA and the patients you've seen and what you know from the literature, say more about the prototypical cognitive symptoms that that tend to come out.

Wendy Kelso 14:25

So Ryan, seeing people with PCA over a number of years, probably one of the early features that people don't always recognize is very significant anxiety. And it's not a cognitive feature. But the reason that they have such significant anxiety is because their cognitive changes are subtle, and they're quite hard to explain. So when people have visuospatial difficulties, unlike memory difficulties, which are easy to explain to people, they know something's wrong, and they often can't articulate the type of changes, and they also sound unusual. So a lot of people with PCA are often diagnosed with a functional neurological disorder, or they're considered to have a functional disorder to begin with, particularly because some of the visuospatial changes and when they're copying things such as a clock face or, or even writing letters, people can't actually believe that they could be so impaired and actually still present very well in interview. So those symptoms are hidden, as you say. So they're more cortically blind, but their visual fields are essentially normal. The classic difficulties, I suppose, what we've always been seen in terms of Balint's syndrome, so simultanagnosia, optic ataxia, and ocular apraxia. So difficulties actually recognizing, so, more than one thing at a time, so an inability to perceive the entire of a visual scene, rather than just focusing on an individual item in simultanagnosia.

With optic ataxia, people often have difficulties. For example, if you're examining them, if you get them to fixate on your nose, and then try and actually move their finger to your finger or track your finger sort of in the visual fields, they have great difficulty with the eye motor coordination, and ocular motor apraxia means that they have difficulty, so the initiation of sort of horizontal saccades, but they also can't actually locate items in space. So people often, if you're testing them, for example, you might put, they might put something in front of them such as a cup and try and get them to locate and pick it up. And they often overreach or under-reach and actually have difficulties trying to find that object, even though it's right in front of them. Some of the more classic features as well of PCA is difficulties reading, and most people run with PCA, particularly in the early stages are still actually able to read, but they need to be able to reduce the distraction and also tracks so because they get the lines mixed up. And sometimes they miss out parts of words that can be quite disabling. So they often need to use strategies, such as putting a piece of paper above or beyond, you know, on top or below to be able to track the words carefully. Sometimes they have difficulties actually perceiving the letters in the same way. So sometimes it's actually easier with smaller print rather than larger print or the print needs to be changed, or they have difficulties with the color perception. So for example, they might not be able to read green or red on the board and it needs to be in clear black or white writing. Often people have difficulties with calculations, particularly early on in the disease process. And those classic what we call sort of visuospatial and visuoconstruction difficulties are very evident early on in the disease. Some of the more commonly used, neuropsychological tests aren't always as sensitive to PCA. And sometimes you need to use tests that look at

even just basic visual discrimination, because people may do poorly on executive tests. But the actual difficulty is not due to executive dysfunctions, it's due to visual spatial dysfunction. So for example, if you're doing something such as coding where someone has to look up and down and transcribe symbols, you're not really looking at psychomotor speed, you're looking at a visual difficulty in terms of the difficulties completing that task. And the same, for example, with trials. You're actually looking at a visual search difficulty, rather than a processing speed or sort of divided attention difficulty. So you need to go back to some of the more basic assessments of vision, you know, what can they actually perceive? Can they perceive fragmented letters? Can they identify colors? Can they identify shapes, those type of more behavioral neurology assessments are quite useful to then be able to work out what are the tests that you'll be able to use that will actually appropriately assess the domain for what they're meant to be assessed for? If that makes sense.

John Bellone 18:53

It's surprising to me that the ophthalmological examination was normal. Do you have thoughts about why that might have been?

Wendy Kelso 19:00

Yeah, it's often quite common, John, in the beginning for because there's nothing wrong with the actual, I suppose visual pathways, it's more of a cortical blindness that people have in terms of occipital and parietal damage, often that their visual fields are considered full. Look, sometimes there's part neglect or if someone's really a lot of optometrists are not looking for this type of difficulty, I suppose, or even ophthalmologist, and it's rare. So PCA is probably about 5% of all Alzheimer's disease. So it's a pretty rare presentation of the disease. And so people are not, often they perform well on some visual tests, a Snellen chart, for example, if it's reduced enough or they've covered up the other letters, often they can do it individually, identify the letters for example, in terms of just visual acuity, but often they have great difficulty if it's not individualized, but they can't actually say the letters that clearly.

John Bellone 20:03

I wonder if neuro-ophthalmologists maybe then would be more attuned to this than just a general ophthalmologist, you'd think that they would be on the lookout for it, but neuro-ophthalmologists would really be the specialists that would be most relevant here.

Wendy Kelso 20:19

Yeah, that's right, John. And a lot of the patients that we see with PCA have seen lots of had lots of optometry ophthalmology investigations for many years before because they describe unusual symptoms such as misperceiving objects or Halo phenomenon where they may see an image and then the image goes away. But then they still see that image after time. Or they look at an object and they've actually misperceived it. So they thought, for example, it was a cat and it was actually a dog or a rodent that they've seen some of the image and

they've tried to identify, you know, use semantic knowledge and trying to identify what it is, but they haven't received the entire image and hence have incorrectly identified what it is.

Ryan Van Patten 21:07

Just to emphasize a few things that are typically spared in PCA that we are used to seeing impaired: things like episodic memory, executive functions, behavioral changes, these things are not common, at least in early PCA, and this was borne out with your patient, Wendy. Language skills would fit in that bucket as well. So language, learning and memory, especially the Alzheimer's disease traditional memory profile, that tends not to be the case. We don't see something like bvFTD there might be anxiety as with your patient, and if we try to imagine what this might feel like for somebody to be so visually, to have such visual distortions in the environment on an escalator, for example, the the anxiety makes sense, but there aren't frank behavioral changes: disinhibition impulsivity, extreme apathy, compulsive behavior, like we see in frontally mediated syndromes.

Wendy Kelso 22:06

That's right, Ryan, often took quite late on the disease process, personality is preserved. So the person is exactly the same as they were, apart from heightened anxiety, which is markedly different, for example, for people that have bvFTD or another, for example, corticobasal syndrome, which is often a differential for posterior cortical atrophy or Lewy Body disease. So, in corticobasal syndrome, often, due to the involvement of the frontal lobes, there's often personality change over time, which is differentiates it from PCA.

John Bellone 22:46

Unlike other neurodegenerative conditions, as the pathology spreads past the occipital and parietal lobe, we could expect other symptoms to come on board that are not classic PCA symptoms, of course.

Wendy Kelso 23:01

Yeah, John. And I think as the disease progresses, and there's more burden of pathology in the other cortices of the brain tends to look more particularly in the middle to late stages, like typical amnestic Alzheimer's disease, where you have some executive difficulties, obviously, memory eventually goes as well over time. Often people with PCA actually a little bit earlier than the memory changes, become a little bit less fluent and have significant word-finding difficulties. And they also have difficulties, you know, with speed and other things over time where it becomes more of a global dementia process. And I think, while amyloid PET is really useful to determine whether PCA is caused by Alzheimer's pathology, it's not very specific, so people with an amnestic form of Alzheimer's disease and also a posterior variant or a language variant, amyloid PET actually looks quite similar in each of these conditions. But if you look at tau imaging, often there is more of a cortical predominance in the occipital and parietal lobes in PCA.

John Bellone 24:11

Yeah, that was interesting that you had MRI, SPECT, and amyloid PET on this patient and it all converged more or less. I'm curious if we could just wrap up by talking about recommendations that you made to your patient other than maybe seeing a neuro ophthalmologist instead of just the general ophthalmologists. What were your recommendations for this patient? And just generally speaking for PCA?

Wendy Kelso 24:35

Yeah, so it depends a little bit on the stages of PCA. I think as people progress and have more visual difficulties we often recommend that people get involved with an organization in Australia called Vision Australia, which is essentially like the Blind Society, I suppose, which may happen in America. So even though that they're not specifically set up for people that have PCA and the type of visual difficulties are quite different if someone has actually got vision impairment, or blindness, some of this, some of the different types of techniques, or suggestions can be quite useful for people with PCA. And I think some of the difficulties people with PCA have is that they might be talking to someone, for example, but they can't locate where that person is in space. So they're not actually talking, they're not looking at the person because they can't actually work out where the person is. So having something such as a cane or even a science and they've got vision impairment can be quite useful in sort of the moderate to later stages of the disease. In terms of function at home, we often give recommendations in terms of things such as increasing lighting, making sure that there's greater differentiation between, for example, if you're sitting down to a meal, not having a white plate on a white placemat on a white table, where someone won't be able to identify where the plate is or where the placemat is or where the cup is, for example. So simple things, such as having light switches that might have a little bit of velcro or something on it that people can see. So they can identify where the lights switch is, is, or using something such as Alexa, you know, to turn on lights, I'm using a verbal cue. We often provide recommendations for people if they enjoy reading, but they're no longer able to read using audio books. And when we're giving people feedback, John, we often would record the feedback in an audio format, so they can play back the feedback session to themselves rather than sending them written recommendations. And I think because I've been involved in with posterior cortical atrophy support groups, one of the great difficulty is this, if you've got people with PCA in the group and family members is often people use PowerPoint or you know, Google Slides as a way to try and present information. Well, people with PCA can't read, so that's no use in terms of being able to provide education. So you need to think of other ways, verbally-mediated ways that you can provide the information that they need in a way that they can sort of register and take it on board as best you can.

Ryan Van Patten 27:15

Something that comes to mind for me when thinking about PCA, especially at the very earliest stages is just, I think that our culture in between first century and the Western Hemisphere and how language reliant we are, where if somebody has a language based syndrome, PPA for example, they start having word finding problems, or they have trouble expressing themselves and understanding that it's going to be evident pretty quickly for most people in our culture. Whereas visual spatial skills we don't rely on as much as we rely on language, we're

using GPS, we have cars, we get people to drive us surround there's just not a lot like in my life. And I as a psychologist, especially language oriented, but visual spatial skills in navigation just aren't as big of a, there's not as big of a demand, for me. But that's not the way it is all over the world. So we should think about how this might look in other cultures. And then of course, as the disease progresses, as I've been talking about, and visual spatial issues become more extreme, the impairments become more severe than certainly it comes out in everyday life. Driving, of course, comes to mind, but in other ways as well. So I just want to think about this in a broader and more holistic way. And we talk and think so much about language, memory and executive problems, and rightly so that we see a lot of patients who have them. But it's, it's interesting and important for us to think about the back of the brain. So this episode is, is all about the posterior parts of the brain. And thank you, Wendy for talking through all that. Before we let you go here, we'd like to ask you a few of our bonus questions, which are just two quick questions about the field of neuropsychology more generally, they don't need to necessarily pertain to PCA. Of course they could but entirely up to you which direction you go. So the first question is, if you can improve one thing about the field of neuropsychology, what would that be?

Wendy Kelso 29:17

Oh, big question, Ryan. I probably should have thought about this a little bit more beforehand. I think often with neuropsychology, particularly in the earliest stages of people's career, they're often very focused on the assessment and diagnosis and the testing, without having full realization for what that means for people in real life. And I think as a profession, we have to get much better at actually putting ourselves in the patient's/in the family's, shoes about what does this actually mean for their everyday functioning? What must it be like to be diagnosed with neurodegenerative illness at a young age, for example, and potentially lose your marriage, have a different relationship with your children, lose your job, and have an honest that is, for example, like PCA, quite frightening when you have difficulties navigating around and become disabled very, very quickly, even though you still got preserved insight, and other cognitive features are still very preserved. So I think that will be something that I would probably try and change is the education I suppose for other neuro psychologists about just how important it is both to give really good feedback sessions, and also post diagnostic support. Because often, I think, neuropsychologists think, well, we've got the diagnosis, right? And that's important. But really, for the people, a diagnosis without follow up is actually quite traumatic. And we talk to lots of people that they feel particularly in the dementia space, unlike cancers where they're given a diagnosis, there's very limited treatment options. And they feel that they're left flailing a little bit with not very much hope. And so I think there's been a lot more work done in the post diagnostic space recently, which I think is fantastic. But I think that that needs to increase considerably for people living with these type of progressive neurodegenerative conditions.

John Bellone 31:18

Yeah, excellent. The last question for you is, what is one bit of advice that you wish someone had told you when you were training? Or maybe somebody did tell you that really made a difference, just an actionable step that trainees can take that they might not have thought of?

Wendy Kelso 31:34

I think when I look back on my training, the people that had the greatest influence were ones that really put themselves in the patient's shoes and tried to get a sense of what it might be like for them. Yeah. And I think what I wish I'd realized at the time was that you need to be a psychologist first, and then a neuropsychologist, second. And I think in the training, we focus so much on the tests and the brain and, and the neural pathways rather than what it might like be for the human condition. And I think you need to have excellent psychological skills before you have neuropsychological skills to be able to relate to someone, establish rapport quickly, or get an accurate history. Because if you don't have good rapport, then you don't actually get the full picture. And therefore you also can set up a therapeutic intervention at the end of the assessment, where you have trust with the patient in the family. And then you can tailor and use that trust to really support the patient ongoing once that you've got the diagnosis and often have an ongoing relationship with the family over time as the illness continuous and progresses.

John Bellone 32:45

We are in full agreement with you.

Ryan Van Patten 32:48

Great answers. And thank you so much for your time, Wendy. This was wonderful.

Wendy Kelso 32:52

Thanks so much, Ryan. Thanks so much, John. It's been a real privilege and a delight to talk from Australia.

Exit Music 32:58

John Bellone 33:02

Well, that does it for our conversation with Wendy. If you'd like to support what we're doing here, please leave us a rating on whatever podcast app you're listening to this on. And as always, thanks so much for listening and join us next time as we continue to navigate the brain and behavior. The navigating neuropsychology podcasts and all the linked content is intended for general educational purposes only, and does not constitute the practice of psychology or any other professional healthcare advice and services.

Ryan Van Patten 33:53

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