PEER REVIEW HISTORY

ARTICLE DETAILS

TITLE (PROVISIONAL)	A qualitative, exploratory pilot study, to investigate how people living with posterior cortical atrophy, their carers and clinicians experience tests used to assess vision.
AUTHORS	Bowen, Michael; Zutshi, Harry; Cordiner, Martin; Crutch, Sebastian; Shakespeare, Tim

VERSION 1 – REVIEW

REVIEWER	Lin SHI The Chinese University of Hong Kong, Hong Kong, China
REVIEW RETURNED	21-Dec-2017

GENERAL COMMENTS	This paper used 3 samples to carry out a pilot study about PCA and was trying to find how different tests are used to assess vision for people with PCA and whether a more effective test exists among them. With efforts of the researchers, some conclusions were drawn in the study. They found that simple and short tests were better and more effective than more subjective ones. Also, during the test, patient's vigour should be an important factor that might affect the result. The largest limit of the study is the small number of samples involved. In the future, reseachers can recruit more age and gender matched individuals with different diseases, e.g., dementia, as well as control group for more comprehensive comparisons.

REVIEWER	Elena Salobrar-Garcia Instituto de Investigaciones Oftalmológicas Ramón Castroviejo. Facultad de Medicina. Universidad Complutense de Madrid
REVIEW RETURNED	30-Dec-2017

GENERAL COMMENTS	1. Purpose Posterior cortical atrophy (PCA) is a cortical neurodegenerative disease that shares physiopathological similarities with Alzheimer's disease (AD), although each one has a different clinical expression. Nowadays, the existence of a dilemma as to whether PCA is a subtype of AD or a different disease is widely recognised, despite both diseases being seemingly distinguishable through a complete neuropsychological evaluation, and despite the data obtained from such evaluation concluding that patients with PCA show specific needs and difficulties that require special attention and intervention. After performing a lumbar puncture, PCA and AD present with the same clinical findings (reduced level of β-amyloid 42 and increased levels of tau protein and phosphorylated tau in the cerebrospinal fluid). Likewise, in a postmortem study, patients of PCA present the same anatomopathological findings usually found in AD patients - the accumulation of neurofibrillary tangles and senile plaques.

	However, studies using PET, SPECT or MRI show that the neurodegeneration pattern in both diseases differs– while degeneration in AD begins in the temporomedial region, degeneration in PCA begins in the occipitoparietal cortex and affects the dorsal and ventral pathways of visual processing. These two different neurodegeneration patterns bring with them two differentiated clinical expressions, and, therefore, considering PCA as a pathological separate entity from AD could be a valid option. There are, indeed, very few reported cases of this disease. That does not mean that no other cases exist, but that they are not documented or published. This is why publishing single case studies is so important, as they are a valuable source of information for other clinicians, and therefore, the work presented by the authors is of interest. However, for the work to be published some corrections would be required. 2. Methodology 2.1. The study has been carried out by an interdisciplinary team made up of an optometrist, a neurologist and an ophthalmologist, who met regularly to reach an agreement on the examination criteria. The authors should mention the specific tests performed, and the results obtained that led to the diagnosis of PCA. 2.2. There is no mention whatsoever from the authors about what the patients' symptoms are, whether the symptoms of visual disturbances have had an insidious onset, whether the absence of an ophthalmologic pathology to explain the symptoms has been analysed, whether visual defects are disproportionate compared to other cognitive deficits, whether the course of the disease was progressive, whether tests were performed to prove the existence of unilateral or bilateral occipitoparietal or occipitotemporal atrophy, hypometabolism or hypoperfusion. Proving all this is essential to discard the existence of such a rare disease as PCA in neuropsychological tests at a later stage. 2.3. The authors mention they have carried out a series of non-specified optometric tests. It would be advisable to know the visual exploration protocol performed on these patients, by both the optometrist and the ophthalmologist. 2.4. The authors evaluate the visual tests as well or badly performed, but do not provide the values obtained from each test. 2.5. No references to fixation and saccades anomalies in the PCA are provided, despite the bibliographic resources available that support the alteration of these functions in this pathology. This could have affected the results of the tests performed. 3. The discussion about the results lacks a thorough reasoning and bibliographical supporting resources. Statements about the pathology are not referenced, and this is essential. The authors do not mention visual disturbances that are known to be experienced by the patients, such as prosopagnosia, apperceptive visual agnosia, or simultagnosia. There is no mention to which types of visuospatial deficits the patients have — optic ataxia or ocular apraxia. These are all vital signs when evaluating a patient's disability, whose importance is highlighted by the authors when stating that the prevalence of visual impairment was found to be more than 2.5 hours in residential settings. 4. Conclusions From the tests carried out to obtain the results of the current study and the explanations given by the authors, it cannot be concluded

	that ‘a simple test which compared full and fragmented images or letters was agreed to be the test that provided the clearest evidence of PCA, or of symptoms of other cortical vision problems, as patients could identify the full image but nor the fragmented one.’ 5. References The authors should do a more comprehensive literature review and discuss the results, as the current bibliography is insufficient and has little relation to visual recognition in PCA.

REVIEWER	Jeffrey Phillips University of Pennsylvania United States of America
REVIEW RETURNED	13-Feb-2018

GENERAL COMMENTS	Bowen et al. (2017) report several observations from patients with posterior cortical atrophy as well as their clinicians in response to competing a number of vision assessments. The authors touch upon multiple topics, including qualitative descriptions of patients' performance, patient fatigue and frustration in response to the tests, the difficulty of diagnosing PCA, and the complications that these patients face in obtaining an accurate diagnosis for a rare syndrome. The reported results--in particular, quotes from patients and caregivers--are fascinating and may help investigators empathize with their patients. However, the manuscript in its current form has contradictory statements about the objectives of the study, and I would urge the authors to draw upon what must be a rich dataset of visual performance measures and transcribed patient-clinician interactions to support their observations and interpretations. I note first that one of the investigators, Harry Zutshi, is named repeatedly in the Procedure and Author Contribution sections but is not included in the author list. This is an alarming omission--the authors are asked to re-check the author list and properly credit all investigators. Additionally, the manuscript would benefit from major revision to clarify the study objectives. These are variously stated throughout the abstract and manuscript itself as: 1. To describe patients' subjective experience of vision tests. Presumably, such information could provide insight to clinicians about how and why patients fail or succeed on vision tests. 2. To identify vision tests that discriminate between cortical and optical/ocular vision impairments. This is a compelling research goal, but the study design (all 3 cases have PCA; there is no comparison group with optical/ocular deficits) does not seem capable of addressing the question. The abstract makes no further mention of the ability of tests/screens to discriminate between causes of vision impairment, although the section "Learning from the tests" notes that physicians agreed upon identification of fragmented images (e.g., letters) as a sensitive task for identifying PCA. 3. To assess the feasibility of administering multiple tests to PCA patients. This seems like a purely logistical question whose scientific value is questionable--it's more important to know what information the tests convey than to know whether PCA patients can complete them. Nevertheless, this is the only conclusion noted in the Abstract. I would suggest that the most compelling of these objectives is the

	second, followed closely by the first. As noted, a series of 3 PCA cases is not sufficient on its own to address this question, unless the authors can provide more detailed description/analysis of how the tests administered distinguish within each patient between performance deficits due to cortical vs. optical/ocular issues. Such a close analysis of visual assessment data would hold significant interest for cognitive neurologists and eye care professionals alike. If the authors are in fact able to make such within-patient distinctions, I recommend highlighting such results as the simplest and most scientifically valuable route of revision. If the current dataset does not allow the authors to effectively distinguish between cortical vs. optical/ocular deficits, there is still considerable value in revising the manuscript to highlight patients' subjective experiences of each assessment. This topic is addressed in general terms: for example, they report, "Other optometric, ophthalmic and neurological tests were generally effective". While the current study is qualitative in nature, there is still considerable room for the authors to provide more detailed methodological description and results. In regards to the previous quote, it would help to know which tests are referenced and how effectiveness was determined. Similarly, the authors note that "more subjective tests such as colour vision, depth perception and visual acuity were more of a struggle for patients." It is unclear what criteria the authors are using to classify tests as objective vs. subjective, or how the authors determined that patients struggled with certain tests. In describing the role of study partners ("The test experience", last paragraph), the authors provide a level of interpretation that approaches editorializing and is not supported by reported results, e.g., "Patients could turn to their partners for assurance during the tests, which could be given simply as a nod of encouragement or the prompt of a correct word." Rather than discussing hypothetical actions on the patients' part, it would be more appropriate to report how often patients actually did receive prompts or encouragement from study partners, or specific instances of their doing so. To increase the usefulness of this study to clinicians and researchers, a complete list of optometric, ophthalmological, and neurological tests administered should be included. Given the small sample size, reporting of individual data is highly feasible, and I request that the authors provide results for each assessment as well as individual demographic data for each patient. Relatedly, Figure 1 does not distinguish between themes expressed by clinicians vs. patients. This is an important distinction if the authors wish to evaluate patients' subjective experience of vision tests. I ask the authors to please revise this figure to separately present themes raised by clinicians and patients. Noting overlap in these themes may also be helpful. The text also includes multiple observations that provide interesting insight into the motivations of PCA patients and their caregivers as well as their interactions with the health care system but have little relation to the study aims. For example, the authors write: "Partners and patients were vocal in their commitment to research projects such as this one", and elsewhere, "Patients thought that they fell between different clinical disciplines, going from one to another with no definitive diagnosis." This content, while useful for understanding and empathizing with patients, does not address the stated focus, patients' subjective experience of vision tests. I would suggest it is

	more appropriate for a separate article. Finally, I add some minor observations: Given the multidisciplinary approach and, consequently, the likely diversity of backgrounds among readers, the authors should describe and, when appropriate, provide citations for field-specific terms such as a a Snellen letter chart and an Amsler Grid. The authors alternately state that participants were told that the study purpose was "to gather data about the experience of having vision/eye health assessed by a range of clinicians", and that participants were unaware that the study focus included "how tests were experienced". Given the first statement, the second appears inaccurate; please clarify. The callout boxes with patient and clinician quotes are not integrated with the article--it is unclear which of the authors' observations or interpretations each is meant to support. The authors have more work to do in not just selecting quotes of interest, but incorporating them into the text to support interpretations. In general, the text needs more support in the form of reference citations. For example, in the fourth paragraph of the introduction ("Purpose"), the authors should provide citations for each of the difficulties that they claim PCA patients experience on vision tests. Some references in the reference list are not included in the text (including Kitzinger, 1995; Pelak et al., 2011; Tong et al., 2007). The manuscript would benefit from proofreading. For example, the authors alternately refer to "Alzheimer's disease" and "Alzheimer's Disease", and "Too long and the patient may become too tired to continue without a break" is a sentence fragment.

REVIEWER	Joost Heutink Royal Dutch Visio Centre of Expertise for blind and partially sighted people Department of Knowledge, Expertise & Innovation Amersfoortsestraatweg 180 1272 RR Huizen The Netherlands University of Groningen Faculty of Behavioural and Social Sciences Department of Clinical and Developmental Neuropsychology
REVIEW RETURNED	02-Apr-2018

GENERAL COMMENTS	-The topic of this paper is very interesting! Assessment of visual and visuoperceptual functions is an important element of the diagnostic process of PCA, since (higher-order) visual disorders are a hallmark of the disease. Especially when the disease may be progressed beyond the initial stage it can be difficult to establish the exact nature of the visual disorders. Low vision assessment may prove to be difficult (patients not knowing where to look on a Snellen chart, having difficulties directing their gaze, etc), visual field examination may be compromised by (spatial) attentional deficits and many neuropsychological tests (either measuring visual perception or other cognitive domains) may be too difficult for patients to complete. Apart from these validity and reliability-related issues, testing may be very disheartening for patients and, if present, their family. Knowing which tests are relatively insensitive to the validity issues and which allow a clinician to discriminate between lower-

	order and higher-order visual functions and to assess the severity of the visual disorders and yet are as patient-friendly as possible is very important. -The paper could do with quite some improvement. However, I would encourage the authors to resubmit. This is an interesting and important issue. -This paper describes what I would rather call a small exploratory pilot in three patients, rather than a study. There is a danger of over interpreting the results of this limited pilot. I feel the authors are in danger of overplaying their hand a little. The scientific and clinical reputation of the authors is without doubt. However, the paper would benefit from avoiding even the suggestion of arguments from authority. Limiting the conclusions and recommendations to what can be concluded from this particular pilot would make this paper a bit more modest but of better quality. -The focus of the study could be described more clearly. It appears to change a number of times in this paper. -It would help a lot if you would name (and possibly describe) the different tests used during assessment. Did all three professionals assess the same functions? Normally, assessment of visual functions will at least consist of refraction, measurement of visual acuity (and perhaps reading acuity, and perhaps at different lighting conditions), assessment of oculomotor function, standardised visual field perimetry and measurement of contrast sensitivity. If necessary and depending on the information available from (the referring?) ophthalmologist, full ophthalmological assessment may be performed. Assessment of neuropsychological functions would probably consist of the Mini Mental State Examination or an equivalent, a memory test, tests for visual (in)attention, tests for visual perception. In many countries a neurologist would not use neuropsychological tests, but rather perform a (full) neurological examination. -The patients participating in this study may have had rather progressed PCA. This is a potential bias in the study. Another option would have been select patients at an early stage of the disease. This would have given a better impression of which tests can be used to detect PCA.

	- In general, the procedure could be described much clearer. For instance, differentiate between the preparation of the experiment, the testing (which tests, by whom), the focus group, the post-testing interviews. -I struggled finding out what was precisely assessed by the three professionals? Different terms are used in this paper. The paper would benefit from clear terminology. In case of PCA I would try to avoid the word eye test. Many patients (and their relatives) find it hard to understand that there may be little wrong with their eyes. -I think it is good suggestion to to include tests that assess holistic perception, such as Incomplete Letters, when assessing patients with (probable) PCA. However, in this pilot this particular test appeared to be to be the most effective in only three patients. I had a look at an old master thesis of one of my students, analysing test performance of 19 patients with PCA we assessed in our multidisciplinary team in the North of the Netherlands. As you can see from the table below, Incomplete Letters of the VOSP appears to be very sensitive. But so do Position Discrimination, and to a slightly lesser extent Dot Counting, Progressive Silhouettes. Please also note that the sensitivity of a particular subtest may be high, but this doesn't mean the selectivity of a test is high as well. Also, some of my patients were not able to perform any of the subtests of the VOSP. **[Please note that I share this information under embargo / in confidence. Do not use, copy or share it without my explicit approval (j.h.c.heutink@rug.nl)]**

Patient	Performances on neuropsychological tests									
	CST	MMST	VOSP							
			LETT	SIL	OBJ	PROG	DOT	POS	LOC	CUBE
1		--	-	-	-	-	-	-	-	-
2	+		-	-	-	-	-	-	-	-
4	+			-				-		
5	+	-	-	-	-	+	+	-	+	-
6			-							
7				-		-	-	-	-	-
8	+		-	-	-	-	-	-	-	-
10		-	+	+	-	+	+	-	+	+
12		-	-	-			-	-	-	-
13		-	-	+	+	-	-	-	-	+
14	-									
15	+		-	+	+	-	-	-	-	-
16	+									
17	+	--								
18		--								
19	-	--								

Note: VOSP: LETT= Incomplete Letters; SIL= Silhouettes; OBJ= Object Decision; PROG= Progressive Silhouettes; DOT= Dot Counting; POS= Position Discrimination; LOC= Number Location; CUBE= Cube Analysis

VERSION 1 – AUTHOR RESPONSE

Reviewer Comment	Author response
The topic of this paper is very interesting! Assessment of visual and visio-perceptual functions is an important element of the diagnostic process of PCA, since (higher-order) visual disorders are a hallmark of the disease. Especially when the disease may be progressed beyond the initial stage it can be difficult to establish the exact nature of the visual disorders. Low vision assessment may prove to be difficult (patients not knowing where to look on a Snellen chart, having difficulties directing their gaze, etc), visual field examination may be compromised by (spatial) attentional deficits and many neuropsychological tests (either measuring visual perception or other cognitive domains) may be too difficult for patients to complete. Apart from these validity and reliability-related issues, testing may be very disheartening for patients and, if present, their family. Knowing which tests are relatively insensitive to the validity issues and which allow a clinician to discriminate between lower-order and higher-order visual functions and to assess the severity of the visual disorders and yet are as patient-friendly as possible is very important.	We would like to thank the review for these comments. JH's comments about low vision assessment agree with the views that led to the initiation of this explorative pilot study. At the time of initiating the project the team did not have funding for a full quantitative investigation to establish the relative sensitivity and specificity of an exhaustive or extensive list of vision assessment tests. There was scope within the resources and funding available to support the explorative pilot study to investigated qualitatively the experiences of people living with PCA, their family members and health professionals in relation to some of the most common tests used to assess vision in primary and secondary care eye health settings, and in neurology settings in secondary care. The objective in addition to gathering qualitative data relating to how tests to assess vision are experienced, this pilot study aimed to explore the viability of research in this area - to establish the key factors enabling or limiting such research with people living with PCA.

	We did not seek to address this aspect of the project in this paper.
The paper could do with quite some improvement. However, I would encourage the authors to resubmit. This is an interesting and important issue.	We appreciate this comment, and the detailed and helpful further comments provided - we have sought to address each of these fully in the revised draft.
This paper describes what I would rather call a small exploratory pilot in three patients, rather than a study. There is a danger of over interpreting the results of this limited pilot. I feel the authors are in danger of overplaying their hand a little. The scientific and clinical reputation of the authors is without doubt. However, the paper would benefit from avoiding even the suggestion of arguments from authority. Limiting the conclusions and recommendations to what can be concluded from this particular pilot would make this paper a bit more modest but of better quality.	This is a helpful overview - thank you. We have sought to revise the paper to more clearly reflect the scope and scale of the project, and its structure as a piece of explorative, pilot research.
The focus of the study could be described more clearly. It appears to change a number of times in this paper.	We have edited the paper to try and address this point. We were clear about the objectives: to gather qualitative data relating to how people living with PCA, their family member carers and health professionals experience various eye health assessments. A further objective was to gather data about the viability of conducting such research with people living with PCA, with a view to informing future larger scale, quantitative research to explore the
It would help a lot if you would name (and possibly describe) the different tests used during assessment. Did all three professionals assess the same functions? Normally, assessment of visual functions will at least consist of refraction, measurement of visual acuity (and perhaps reading acuity, and perhaps at different lighting conditions), assessment of oculomotor function, standardised visual field perimetry and measurement of contrast sensitivity. If necessary and depending on the information available from (the referring?) ophthalmologist, full ophthalmological assessment may be performed. Assessment of neuropsychological functions would probably consist of the Mini Mental State Examination or an equivalent, a memory test, tests for visual (in)attention, tests for visual perception. In many countries a neurologist would not use neuropsychological tests, but rather perform a (full) neurological examination.	We have added some additional information about the vision assessments used. Since we were not in this small study seeking to explore the experiences of neurology assessments, we asked the neurologist to focus on assessing vision / visio-perceptual functions rather than conducting a complete neurological assessment. In this pilot the objective was not to precisely recreate the experience of each of the examination settings
The patients participating in this study may have had rather progressed PCA. This is a potential bias in the study. Another option would have been select patients at an early stage of the disease. This would have given a better	Thank you for this comment - we agree that this was a limitation of the study. We worked with the UCL PCA support group to identify volunteers who were both willing to take part, and available to participate in the day of testing. Younger

impression of which tests can be used to detect PCA.	participants, earlier in the progression of their PCA would be important to include in any future, larger scale quantitative research to evaluate the sensitivity / specificity of vision assessment tests / tools.
In general, the procedure could be described much clearer. For instance, differentiate between the preparation of the experiment, the testing (which tests, by whom), the focus group, the post-testing interviews.	Thank you for this helpful comment - we have edited the paper to add additionaldetails of the procedures.
I struggled finding out what was precisely assessed by the three professionals? Different terms are used in this paper. The paper would benefit from clear terminology. In case of PCA I would try to avoid the word eye test. Many patients (and their relatives) find it hard to understand that there may be little wrong with their eyes.	In the UK the terms 'sight test' , 'eye examination' and 'eye test' are commonly used to refer to vision assessments, especially in primary care. We have sought to edit the paper to use the term 'vision assessment'.
I think it is good suggestion to include tests that assess holistic perception, such as Incomplete Letters, when assessing patients with (probable) PCA. However, in this pilot this particular test appeared to be to be the most effective in only three patients. I had a look at an old master thesis of one of my students, analysing test performance of 19 patients with PCA we assessed in our multidisciplinary team in the North of the Netherlands. As you can see from the table below, Incomplete Letters of the VOSP appears to be very sensitive. But so do Position Discrimination, and to a slightly lesser extent Dot Counting, Progressive Silhouettes. Please also note that the sensitivity of a particular subtest may be high, but this doesn't mean the selectivity of a test is high as well. Also, some of my patients were not able to perform any of the subtests of the VOSP.	This comment was very helpful - especially with the additional background from the master's thesis work. As previously noted, it was beyond the scope and design of this exploratory pilot study to establish the relative sensitivity and specificity of individual tests in a quantitative manner that could be regarded as valid. We were very much interested in gathering informative qualitative data about the participants' experiences of the various tests. The reporting of the fragmented letters as having emerged as the 'best' test for supporting health professionals to distinguish between ocular / optical causes of visual impairment and cortical causes.
Comments within the draft	
P2 - Line 11: 'how are various tests used to assess vision…' – JH comment - Not really addressed in this study "how are tests used?" is a question too much open to interpretation.	We wonder if there is a confusion relating to the use of the phrase 'how are various tests used to assess vision experienced by...' The team wondered if the reviewer had focused on 'how are tests used to assess vision' within this phrase, rather than the complete intended sense which is not to explore how the tests are used to assess vision, but how such vision assessment tests are experienced by those being tested and those administering the tests? We have revised the text to try to address this point.

P2 - Line 12: JH comment – “assess vision not clearly explained in this paper what is really assessed.	Wehave edited the paper to clarify which tests were available and used in the various assessments.
P2 - Line 24: ‘Simple short tests were thought to be more effective than more subjective tests…’ JH comment – both subjective and objective tests can be simple and short. Unclear which tests and who (which professional) is testing what.	Thank you for raising this - we have amended the text to improve the clarity of this section.
P2 - Line 26: ‘Patients and carers made clear the importance of early identification of PCA…’ JH comment – was this part of the research question?	Yes - the project aimed to gather qualitative data relating to how people living with PCA and their family members / carers experienced tests used to assess vision, but their general experience of eye health systems was not excluded.
P2 - Line 27: ‘…current levels of understanding of the condition amongst many health professions involved was sometimes preventing this [early detection].’ JH comment - good point but this may not be very relevant to this study	The study aimed to explore health professionals' experiences of administering tests but did not exclude their views of current systemic issues relating to eye health systems capacity to address PCA.
P2 - Line 31: ‘….evidence that there are suitable eye examination tests that people with dementia can engage with and complete.’ JHcomment – “people with dementia” meaning all forms of dementia or primary PCA?	We have edited this to clarify the intended meaning.
P2 - Line 31: JH comment - unclear what is meant: low vision assessment, assessment of visual functions, assessment of visio-perceptual functions?	Each professional was asked to approach each assessment as they would as closely as possible to their usual practice. These broadly equated to: optometrist - primary care General Ophthalmic Services sight test; ophthalmologist - general secondary care hospital eye service general referral (refraction clinic) vision assessment; neurologist - the visual perceptual elements of a routine neurological examination.
P2 - Line 39: ‘Project might have benefitted from a wider range of screening tests being used…’ JH comment – unclear which tests were used in this study.	Additional detail has been added to address this point.
P2 - Line 47: ‘Multidisciplinary approach, incorporating optometric, ophthalmological and neurological screening tests.’ JH comment – unclear which tests and who (which professional) is testing what.	Additional detail has been added.
P3 – Lines 5 / 6: ‘pilot data from people with the posterior cortical atrophy form of dementia and eye health professionals’. JH comment – unclear terminology	We were not clear what the reviewer's specific concern was regarding clarity of terminology?
P3 – Line 9: ‘with people with posterior cortical atrophy, and to be guided by patient and practitioner experience in beginning’. JH comment - Purpose could be addressed more clearly. This was a pilot to assess which tests would be feasible to use in a larger PCA study? Perspectives (according to this paragraph) are	We have sought to more clearly set out the objectives for this project in therevised paper.

technical, administrative feasibility and patient experience (fatigue, burden)	
P3 – Line 11: 'home in on the most suitable tests to investigate within a larger project.' JH comment - maybe add why it is important to address these research questions?	We have added some additional information to explain why the research is of potential value / importance.
P 3 – Lines 15/16: 'It is most commonly caused by Alzheimer's Disease, although may also be caused by dementia with Lewy bodies, corticobasal degeneration or Creutzfeldt–Jakob disease.' JH comment - better add a reference here. e.g. Crutch et al., 2012; Zakzanis & Boulos, 2001	Thank you for this suggestion - we have included additional relevant references in the revised draft.
P3 – Lines 19/20: First described in 1988, consensus criteria for PCA have only recently been agreed (Crutch et al., 2017) and diagnosis is often delayed or absent. JH comment - add something more about the criteria, linking this to the important of visual assessment and this study? (necessary for one of the core features) JH comment - Is a different problem. Better to address in a different sentence.	We have revised the draft to address these points.
P3 – Lines 22/23: 'Most Alzheimer's disease cases appear in people over 65, but PCA tends to occur between 50 and 65.' JH comment - reference needed here e.g. Galton, Patterson, Xuereb, & Hodges, 2000; Crutch et al., 2012	Thank you for these suggestions - we have added references as requested.
P3 – Line 32:'They may also struggle with excessive visual crowding in their central vision, resulting in difficulty reading letters surrounded…' JH comment - added to that: impaired spatial cognition (Balint's syndrome)	We have added additional detail to address this point.
P3 – Line 36/37: 'The complexities of both diagnosing PCA and ascertaining the best sight possible for people with the condition presented an opportunity…' JH comment - best sight? What is meant by that? Visual acuity, best correction? Better to choose well-defined terminology for different types of visual assessment and use that consequently throughout the paper It seems a new research question is introduced here.	The intended meaning was that it may be complicated for optometrists and ophthalmologists to work with people living with PCA to find the most appropriate approach to correcting visual impairment, given the complexities introduced by the cortical visual perceptual symptoms associated with PCA. Wehave adjusted the text to clarify this point.
P3 – Lines 42/43: 'How are the various tests used to assess vision experienced by people living with posterior cortical atrophy?'	We have edited the text to address this point, clarifying that it is both important for optometrists and ophthalmologists to improve their understanding of how people with PCA

JH comment - Paper would benefit if you explain why these are important questions to address.	experience different tests for assessing vision, and also to try to identify the most effective tests to enable optometrists and ophthalmologists to distinguish between ocular and optical cause of visual impairment, and cortical causes.
P3 – Lines 45/46/47: 'Are there particular tests for assessing vision that are more effective at discriminating between cortical vision problems and vision problems related to optical or ocular causes?' JH comment - very important research question! However, this is something different than the research purpose given in the first paragraph. The introduction (and the rest of the paper) would benefit from more focus.	Wehave tried to clarify this point. While the scale and scope of this small study was not intended to be sufficient to validate a specific test, it was intended to provide insights to inform future research, and part of the interest was in whether there were tests that were accessible to people living with PCA, that were also useful in helping eye health professionals discriminate between cortical and optical / ocular visual problems.
P3 – Line 52: 'Vision testing and post-test interviews took place…' JH comment – unclear.	We have edits to try to clarify this point.
P4 – Lines 3/4: 'to analyse selected footage and discuss a schedule'. JH comment – selected by whom?	We have added text clarifying this.
P4 – Lines 9/10: Due to the nature of PCA, the project aimed to include vision testing techniques from several different health care disciplines. JH comment - In terms of severity/progression? In terms of number of symptoms present? In terms of ventral / dorsal / primary visual presentation?	WE have edited the text to clarify the intended meaning for this section.
P4 – Lines 13/14: '…the number of participants to three people with PCA (one male, two female, ages ranged from 67 to 78).' JH comment - A bit odd to give a range for a 'group' of n=3; better give the age of all three participants. JH comment – so presumably rather progresses stage PCA? How did this influence the outcome?	Wehave added the ages of all of the participants. We have added some additional details about the participants. We have addressed the absence of participants with earlier PCA in the limitations.
P4 – Line 16: 'Participants with PCA were given an information sheet with brief details…' JH comment - which probably could not be read by them?	We have added additional detail clarifying how the team addressed consent and ethical considerations. The research team was experienced in working with people living with PCA and dementia, and these aspects of the study were carefully planned and dealt with.
P4 – Line 17: '…of having vision / eye health assessed by a range of clinicians.' JH comment – new terminology. See comments above.	We are not entirely sure what the reviewer's interest / concern was with regard to this comment.

P4 – Lines 21/22: 'Three clinicians took part on the day - an optometrist, a neurologist and an ophthalmologist (one female and two male).' JH comment – with or without experience with testing people with PCA? How much experience? No neuropsychologist involved?	We have added some detail relating to the experience of the clinicians. We did not have a neuropsychologist in the project as it had not proven possible to recruit one, and time limits on the day of visual assessment precluded testing by an additional professional.
P4 – Line 31: Procedure JH comment - In general, procedure could be described much clearer. For instance, differentiate between the preparation of the experiment, the testing (which tests, by whom), the focus group, the post-testing interviews.	Additional details have been added to this section.
P4 – Lilne33: 'Each patient completed three sequential assessments with an optometrist, ophthalmologist and neurologist….' JH comment - in which order? Was there contact between the professionals about their findings? This would normally happen in a multidisciplinary assessment.	We have provided a schedule for the vision assessments.
P4 – Line 39: 'A group schedule of questions / topics for the clinicians' focus group…' JH comment - Was this before or after the testing? how much time in between?	We have clarified that this was after the day of vision assessments. 2 weeks between assessments and focus group.
P4 – Lines 39/40: '…informed by the themes arising during the eye examinations and post-examination interviews…' JH comment - Eye examinations is a vague term. What kind of examination? Three times the same type of assessment? JH comment - How long did each assessment last? A fixed time or till all tests were (more or less) completed?	We have added additional detail to clarify the nature of tests and the equipment available. We have included a schedule for the vision assessment sessions.
P4 – Line 47: '…prompts were used by the interviewer / focus group facilitator to…' JH comment - First time introduced here (maybe I missed this earlier)? Was the focus group facilitator present during the testing? What was her/his role exactly?	The focus group facilitator was not present during the vision assessments, but did conduct the post-assessment interviews.
P4 – Line 49: '…discussion came from the participants and care was taken to use open questions…' JH comment - How was this achieved?	Open questioning is an established qualitative research method. HZ and MB are experienced in qualitative research methods. The interviewer was supported by the development of semi-structured interview scripts prior to the

	assessment day, to inform the post-assessment interviews, without precluding the scope for participants to discuss other issues.
P4 – Line 52: 'Although the patient participants were aware that they were going to have their eyes…' JH comment - They probably did not (just) have their eyes examined. See comments above about terminology/	For the purposes of this study the assessments were limited to vision and eyehealth assessments / examinations. There was some limited cognitive function assessment inherent in some of the visual tests for cognition, but no explicit cognitive assessments (such as MMSE) were used.
P4 – Line 54: '…study being to identify how tests were experienced and whether any tests were particularly…' JH comment - This is a much clearer rationale for the study than the one given earlier in the Introduction! Or is this a new/different objective?	We have sought to clarify this throughout the paper.
P4 – Line 56: '….the clinicians' focus group, were conducted by one of the investigators (HZ) who has…' JH comment - Does not appear to be one the co-authors?	HZ has been added to the author list.
P5 – Line 3: '….extensive experience of qualitative research, interviewing and focus groupfacilitation.' JH comment - This is not a very 'scientific' , since is appears to be an "argument from authority" Maybe better to describe how, not by whom.	We have edited this section.
P5 – Lines 18/19: 'Consolidated Criteria for Reporting Qualitative Research (COREQ) guidelines were followed in the design and reporting of the study.' JH comment - A reference here would be helpful for the naive reader.	We have added a reference.
P5 – Line 23: 'All sight tests / eye examinations and post-examination interviews were video recorded.' JH comment - See earlier comments: Describe the nature of the three examinations performed by the different types of professionals. Avoid using the term eye test.	We have addressed this point in previous edits.
P5 – Lines 30/31: 'Data were analysed by two of the authors (MB and HZ) independently using framework analysis (Pope, Ziebland, Mays, 2000; Glen, Baker and Crabb, 2014) as shown in **Table 1**.' JH comment - HZ is not an author of this paper	HZ added as an author.

(according to the 1st page).	
P5 – Lines 36/37/38: 'Any differences of opinion regarding the relative importance of themes, or the meanings of sentences were discussed until a consensus was reached.' JH comment - Not clear when assessment was regarded as successful. Could you explain?	We were not entirely clear how this comment related to the section of the paper to which it was linked. We were not seeking to quantitatively evaluate the relative success or efficacy of the various tests in this project. We were seeking to gather qualitative evidence relating to how people living with PCA, their family members and various health professionals experienced the assessments (whether being assessed / tested or administering them)...This section dd
P6 – Lines 5 to 9 (Findings): JH comments - Not entirely clear to me how framework analysis helps to answer the questions addressed in this study. Not clear how fig 1 relates to the analysis.	Framework analysis was used to analyse the qualitative data collected in this study. The approach to answering the questions at this stage in this small explorative project was to gather information about the experiences of people living with PCA, their family members and health professionals carrying out the vision assessments, of those assessments. Figure 1 shows the themes and sub-themes identified at the successive stages of the framework analysis.
P6 – Line 39: 'Clinicians reported that it was difficult to take a reliable history because of patient memory…' JH comment - Was taking history part of one (or all three) assessment(s)?	Yes - we have amended the draft to clarify this. Taking medical history would normally form a part of optometry, ophthalmology and neurology assessments, so was included.
P6 – Line 41: 'many variables were significantly less effective with this group of patients. Examples of less successful…' JH comment - "were" reflects a fact. rather: appeared JH comment - Group is a somewhat misleading term, since only three patient participants were tested.	We have revised this section - thank you for this feedback. We have adjusted the text to reflect this and other related comments by the reviewer.
P6 – Line 43: '…and neurological tests were generally effective; however, more subjective tests such as…' JH comment - This type of info (which tests used) should be provided earlier.	We have added this detail earlier in the paper.
P6 – Line 44: '…colour vision, depth perception and visual acuity were more of a struggle for patients…' JHcomment - Not a clear distinction between the function the test is supposed to assess and the way this is done. For instance, colour vision tests such as Ishihara might cause problems because	We did not seek to comment on these details due to the limited scope and scale of the project, and the fact that it was aimed at gathering qualitative rather than quantitative data. The reviewer is absolutely correct that these are important considerations and distinctions, which we would hope to address in future, larger, scale quantitative research aimed at exploring the

of holistic perception, whereas Hue test may cause problems because of spatial problems (such as dorsal simultanagnosia or optic ataxia).	relative efficacy of tests in more depth.
P6 – Line 45: '…neurological test using full and fragmented letters or images (see Figure 2) appeared…' JH comment - This is a neuropsychological test or a test of visual perception. Not a neurological test.	We have edited the paper to refer to neuropsychological tests.
P7 – Line 22: 'Fatigue was definitely a factor by the end of the day and within the test process…' JH comment - Was there a fixed order of testing?	Please see previous comment relating to the sequence of assessments. We have added detail to the paper to clarify that the health professionals were not asked to adhere to a predefined order or set of tests.
P7 – Line 24: '…meant that the time that testing took was significant.' JH comment - This is a rather obvious observation. I can be applied to any patient (with or without PCA, or with AD, PD, stroke etc). Better to state how long each session took?	We agree that to health professionals and researchers who are familiar with PCA and these other conditions might feel that this was obvious, but we hope that this project / paper will be available to a broad range of primary and secondary health professionals, some of whom will be less familiar with PCA, or the other neurological causes of visual problems.
P7 – Line 41: 'Patients recognised that the testing was necessary, but they also found it uncomfortable and emotional at times, as it focused on what they were not able to do' JH comment - Why was testing necessary? Was it an experiment or was is a normalclinical assessment? Were the patients not assessed before? Since it was not their first visit to the hospital... Were the patients helped remembering that they were allowed to quit at any given time? This ethical issue should be addressed in the paper.	The vision assessments were not carried out as part of the participants' normal care / treatment. This comment relates to patient participant comments about the experience of having their vision assessed repeatedly at successive appointments. They were aware that this was an additional assessment solely for research, and that they could stop taking part at any stage of the process. All participants were competent to consent to take part in the research, and were accompanied at all times by a family member.
P8 – Lines 8/9: '…*which are fairly standard eye tests that even a competent ophthalmologist would pick up necessarily that it is a brain disorder,…'* JH comment - Patient and partner comments could be useful, but not sure what they illustrate. I would suggest to give it more focus.	Patient and partner comments were an integral part of the qualitative data that thisproject sought to gather. They are fundamental to the project.
P8 – Lines 16/17: 'They argued that it is important to look at two different aspects, pre-diagnosis and post-diagnosis…' JH comment - Unclear what is meant by this. Aspects of what?	Aspects of their experience of the health care system and eye health within that - prior to diagnosis they all reported having seen eye health professionals who were unclear about their visual symptoms, usually until they found an optometrist with greater awareness of the potential for neurological issues who referred them to secondary care ophthalmology, where many of them had then also experienced delays before finally being referred to a neurology team. Post-diagnosis their experiences were generally

	more positive, with the relevant health professionals understanding that their visual symptoms will have features that are due to cortical deficits rather than solely ocular / optical issues.
P8 – Lines 20/21: 'If it was possible to develop a simple test or series of tests to give an indication that PCA may be involved,…' JHcomment - I do not entirely follow this line of reasoning. I understand how good assessment of visual and perceptual functions may contribute to diagnosing PCA once it is suspected. This study may contribute to that. However, PCA is also known for its insidious onset. Patients may complain about a sore eyes, hallucinations, problems dressing, reading problems, finding things, etc. They are likely to visit their GP first, who may not be familiar with PCA all, let alone suspect this from the early symptoms (which may be clouded by other co-existing problems).	We agree entirely with the reviewer here - the fact that so many people living with PCA report having seen their GP or an optometrist due to the early visual symptoms, and that they experienced delays in getting referred appropriately (i.e. with suspect cortical visual problems / suspect PCA) was a significant factor in setting this project up. This comment came from the professionals who participated, who like the idea that (if feasible - and almost certainly requiring additional further research) developing a relatively simple set of tests to assist / support health professionals - especially primary care optometrists - to identify people who may have cortical aspects of visual impairment earlier and to enable them to refer with a clear query in this area. Neither the clinicians participating in this research, nor the research team are suggesting that diagnosis of PCA could / should occur in primary care general practice or optometry practice settings, or even in secondary care ophthalmology services - just that if there were relative accessible tests that could flag to a clinician that cortical visual problems might be at play, this could be very valuable in reducing the time taken to get people with early PCA to the neurology team in the first place.
P9 – Line 3: 'As a result of this, a key priority for patients and their partners was that diagnostic systems…' JH comment - What is meant by 'diagnostic systems'? Isn't this typically something for a multidisciplinary approach?	We have revised this as the reviewer's comment helped to highlight that this was less clear than if could be.
P9 – Line 26: '…clearest evidence of PCA, or symptoms of other cortical vision problems, as patients could…' JH comment - Probably the most sensitive in this selection of three patients. The incomplete letter test however is not a "test of PCA", but a test to detect visual perceptual disorders.	We agree completely, and have revised the draft to make this clearer. As with the comment above, we were not proposing that the fragmented letter test would enable optometrists or ophthalmologists to diagnose PCA - rather that it might be one of a range of relatively simple to deploy and use tests that aren't currently used by ophthalmologists or optometrists, but which could assist them in identifying people with suspect cortical vision problems, which in turn could facilitate better referrals into neurology for further assessments.
P9 – Lines 30-33: 'For example, one said she could identify a small crumb on the floor but yet not see a glass on the table. One neurological test looked at visual disorientation.	Again, we agree entirely with the reviewer on this point. However, as previously noted, we hope that this paper will be promoted and accessed by a range of health professionals who may be less

The patient was asked to grasp theclinician's finger, but was often unable to do so' JH - I would argue these are not unusual symptoms for people with PCA. In fact, very common for people with Balint's syndrome due to PCA (or bilateral stroke).	familiar with PCA and Balint's syndrome etc.
P9 – Lines 35-37: 'I *ask patients to grab my finger. This can look like a field defect, but it is not. Patients can see the hand and can copy the hand movement, yet cannot locate the finger in space. There is an unusual visual field and visual disorientation.'* JH comment - This comment appears to mixing up visual field problems and disorders of spatial cognition (such as simultanagnosia). People with simultanagnosia may have serious problems performing perimetric tests (and thus appear to have limited visual fields) wheras in fact their visual field may be intact.	Wehave edited this section to clarify this. It should be noted that the quoted section is an excerpt from the professionals' focus group so should not be edited as it a verbatim quote.
P9 – Lines 40/41: 'One clinician noted that it would be useful to include a routine slit lamp investigation with the tests. Another thought that it might be worth trying other field test approaches.' JH comment - why? To exclude what?	We have updated this section with additional details to clarify this.
P9 – Lines 42/43: 'One patient could not see the light at all, while it came and went for another patient.' JH- 'Seeing' is a difficult term in this case. The patient may not have looked in the right direction. Optic ataxia is not uncommon in patients with PCA	We have added some additional text to address this point.
P11 – Lines 51-53: 'This is positive as it provides some further support for the finding that many people living with dementia could complete most of the key elements of a standard sight test (Bowen et al, 2016).' JH comment - See earlier comments.	Noted - thank you. We have sought to address this in the revised draft.
P12 – Line 6: '…test that provided clearest evidence of PCA, or symptoms of other cortical vision problems…' JH comment - was this a goal of the study? Part of the assessment? Do you mean 'clearest evidence of visual perceptual disorders?	We have updated the draft to clarify this point - we did indeed mean the clearest evidence of perceptual disorders as the reviewer suggests.
P12 – Line 9: '…clinicians felt that simple, short tests were more effective than subjective tests.' JH comment - This is not a real contradiction. Subjective versus objective. Simple versus	Agreed - we have revised this section to address this point.

complex. Short versus long.	
P12 – Lines 12/13: ‘A key priority for patients and their partners was that diagnostic systems were in place to enable early identification of PCA.’ JH comment - I thought this was not a goal of this study?	Identifying the experiences and interests of people living with PCA in relation to vision assessment / eye health assessments was part of the study, while this might be stretching that scope slightly, it emerged as a clear consistent priority across the 3 participants.
P12 – Lines 16/17: ‘These concerns were echoed by the clinical professionals who acknowledged the difficulty many would have in making a diagnosis.’ JH comment - May be absolutely true, but was it investigated in this pilot?	It wasn't investigated quantitatively, but it was a theme that emerged from the qualitative data in this project - from patient, carer and clinician participants...
P12 – Lines23/24: ‘Future research should clarify numbers with PCA, establish cross-profession knowledge and skills in this area, and work on further screening tests for PCA…’ JH comment - I agree, but this is not based on this pilot. I would suggest leaving this out the conclusions and perhaps add to the recommendations.	This is included within the conclusions because this specifically emerged as a theme within the qualitative data gathered during the clinician / professional participants' focus group....We are concluding that the professionals in this study felt that these were important issue for future research to address. We have reworded this to reflect this more clearly.
P12 – Lines 34-36: ‘Refine and simplify optometric and ophthalmological tests to make them more effective for patients with PCA or dementia more widely, and undertake research to find out how these work in practice with larger and more varied cohortsof…’ JH comment - Tests of visual percepion are normally in the domain of neuropsychologists, not optomtrists/orthoptists (at least in the Netherlands) My own experience with patients with PCA is in a multidisciplinary team consisting of an ophthalmologist, optomotrist, orthoptist, occupational therapist and neuropsychologist.	We agree with the reviewer on this up to a point. However, while someone who has successfully made it to a neurology or neuropsychology team / or clinician might well be fortunate enough to then have a multidisciplinary team working on their details assessments, it is not the case that everyone with PCA or other visual perceptual deficits / symptoms are readily recognised by these other professionals if they are not used to working closely with neurology colleagues, which many of them will not be. We are not suggesting that optometrists / ophthalmologists could / should be diagnosing PCA - just that with some different tests and additional information they might be able to better identify and refer people with visual perceptual / cortical vision problems early on... We appreciate the reviewer's comment on this recommendation, but note that at present in the UK there is reasonable evidence that many patients with PCA are first referred into secondary care by primary care optometrists, but arrive without a clear flag on their referrals that there is suspicion of neurological / cortico-visual problems. They then end up in HES ophthalmology / optometry led clinics and may not be successfully identified in those settings either.
P12 – Line 40/41: ‘neurologists as part of the research outlined in point 1, and examine their effectiveness in the diagnosis of PCA…’ JH comment - I think it is a good idea always to	We agree entirely with the reviewer's comments - we are not suggesting that this small exploratory project provides sufficient evidence to support promoting this as the definitive test for PCA - but given the simplicity, speed and low

include tests that assess holistic perception. However, this particular test appeared to be to be the most effective in only three patients. But does this mean Incomplete letters discriminates between people with PCA and people without PCA. For that toknow one would need to know both selectivity and sensitivity. See also my comment in the Word document I attached.	cost of making this test available, we believe it is reasonable to do so prior to any further research to further validate the test in the manner the reviewer suggests.
P12 – Lines 55/56: Michael Bowen co-drafted the manuscript with Harry Zutshi and reviewed and approved the final draft for submission. Martin Cordiner re-drafted the manuscript and approved the final…' JH comment - AHA! This is the HZ mentioned earlier. However not mentioned as one of the authors.	Noted HZ added as an author.
JP Bowen et al. (2017) report several observations from patients with posterior cortical atrophy as well as their clinicians in response to competing a number of vision assessments. The authors touch upon multiple topics, including qualitative descriptions of patients' performance, patient fatigue and frustration in response to the tests, the difficulty of diagnosing PCA, and the complications that these patients face in obtaining an accurate diagnosis for a rare syndrome. The reported results--in particular, quotes from patients and caregivers--are fascinating and may help investigators empathize with their patients. However, the manuscript in its current form has contradictory statements about the objectives of the study, and I would urge the authors to draw upon what must be a rich dataset of visual performance measures and transcribed patient-clinician interactions to support their observations and interpretations. I note first that one of the investigators, Harry Zutshi, is named repeatedly in the Procedure and Author Contribution sections but is not included in the author list. This is an alarming omission--the authors are asked to re-check the author list and properly credit all investigators. Additionally, the manuscript would benefit from major revision to clarify the study objectives. These are variously stated throughout the abstract and manuscript itself as: 1. To describe patients' subjective experience of vision tests. Presumably, such information could provide insight to clinicians about how and why patients fail or succeed on vision tests. 2. To identify vision tests that discriminate between cortical and optical/ocular vision	We would like to thank the reviewer for these comments. We accept this feedback, and have made extensive changes to the paper to address this, seeking to clarify the objectives and outcomes for the project. At the time of submission, it had not been possible to contact HZ to gain his formal approval for the final draft of the paper due to health reasons. HZ has now given this approval, and contributed to the review and revision process. We agree that this would be valuable, but have not sought to address this in this paper - we will write this more detailed analysis of the individual tests once the team has had time to complete the additional analysis needed to support this. We accept this point - we have sought to clarify that this project was an explorative pilot project aimed at gathering pilot data, testing the

impairments. This is a compelling research goal, but the study design (all 3 cases have PCA; there is no comparison group with optical/ocular deficits) does not seem capable of addressing the question. The abstract makes no further mention of the ability of tests/screens to discriminate between causes of vision impairment, although the section "Learning from the tests" notes that physicians agreed upon identification of fragmented images (e.g., letters) as a sensitive task for identifying PCA.	feasibility of such research with people living with PCA, and to allow future research to be designed well in terms of the testing / vision assessment process. We have revised the paper to address this point.
3. To assess the feasibility of administering multiple tests to PCA patients. This seems like a purely logistical question whose scientific value is questionable--it's more important to know what information the tests convey than to know whether PCA patients can complete them. Nevertheless, this is the only conclusion noted in the Abstract. I would suggest that the most compelling of these objectives is the second, followed closely by the first. As noted, a series of 3 PCA cases is not sufficient on its own to address this question, unless the authors can provide more detailed description/analysis of how the tests administered distinguish within each patient between performance deficits due to cortical vs. optical/ocular issues. Such a close analysis of visual assessment data would hold significant interest for cognitive neurologists and eye care professionals alike. If the authors are in fact able to make such within-patient distinctions, I recommend highlighting such results as the simplest and most scientifically valuable route of revision.	This is a helpful and encouraging comment - as noted above, we intend to carry out further analyses of the transcripts and the videos of the vision assessment sessions to explore this question, but have not addressed this within the current paper. We have revised the paper to address this point.
If the current dataset does not allow the authors to effectively distinguish between cortical vs. optical/ocular deficits, there is still considerable value in revising the manuscript to highlight patients' subjective experiences of each assessment. This topic is addressed in general terms: for example, they report, "Other optometric, ophthalmic and neurological tests were generally effective". While the current study is qualitative in nature, there is still considerable room for the authors to provide more detailed methodological description and results. In regards to the previous quote, it would help to know which tests are referenced and how effectiveness was determined. Similarly, the authors note that "more subjective tests such as colour vision, depth perception and visual acuity were more of a struggle for patients." It is unclear what criteria the authors are using to classify tests as objective vs. subjective, or how the authors determined that patients struggled with certain tests. In describing the role of study partners ("The test experience", last paragraph), the authors provide a level of	

interpretation that approaches editorializing and is not supported by reported results, e.g., "Patients could turn to their partners for assurance during the tests, which could be given simply as a nod of encouragement or the prompt of a correct word." Rather than discussing hypothetical actions on the patients' part, it would be more appropriate to report how often patients actually did receive prompts or encouragement from study partners, or specific instances of their doing so. To increase the usefulness of this study to clinicians and researchers, a complete list of optometric, ophthalmological, and neurological tests administered should be included. Given the small sample size, reporting of individual data is highly feasible, and I request that the authors provide results for each assessment as well as individual demographic data for each patient. Relatedly, Figure 1 does not distinguish between themes expressed by clinicians vs. patients. This is an important distinction if the authors wish to evaluate patients' subjective experience of vision tests. I ask the authors to please revise this figure to separately present themes raised by clinicians and patients. Noting overlap in these themes may also be helpful. The text also includes multiple observations that provide interesting insight into the motivations of PCA patients andtheir caregivers as well as their interactions with the health care system but have little relation to the study aims. For example, the authors write: "Partners and patients were vocal in their commitment to research projects such as this one", and elsewhere, "Patients thought that they fell between different clinical disciplines, going from one to another with no definitive diagnosis." This content, while useful for understanding and empathizing with patients, does not address the stated focus, patients' subjective experience of vision tests. I would suggest it is more appropriate for a separate article. Finally, I add some minor observations: Given the multidisciplinary approach and, consequently, the likely diversity of backgrounds among readers, the authors should describe and, when appropriate, provide citations for field-specific terms such as a a Snellen letter chart and an Amsler Grid. The authors alternately state that participants were told that the study purpose was "to gather data about the experience of having vision/eye health assessed by a range of clinicians", and that participants were unaware that the study focus	We have added this detail.

included "how tests were experienced". Given the first statement, the second appears inaccurate; please clarify. The callout boxes with patient and clinician quotes are not integrated with the article--it is unclear which of the authors' observations or interpretations each is meant to support. The authors have more work to do in not just selecting quotes of interest, but incorporating them into the text to support interpretations. In general, the text needs more support in the form of reference citations. For example, in the fourth paragraph of the introduction ("Purpose"), the authors should provide citations for each of the difficulties that they claim PCA patients experience on vision tests. Some references in the reference list are not included in the text (including Kitzinger, 1995; Pelak et al., 2011; Tong et al., 2007). The manuscript would benefit from proofreading. For example, the authors alternately refer to "Alzheimer's disease" and "Alzheimer's Disease", and "Too long and the patient may become too tired to continue without a break" is a sentence fragment.	
ESG 1. Purpose Posterior cortical atrophy (PCA) is a cortical neurodegenerative disease that shares physiopathological similarities with Alzheimer's disease (AD), although each one has a different clinical expression. Nowadays, the existence of a dilemma as to whether PCA is a subtype of AD or a different disease is widely recognised, despite both diseases being seemingly distinguishable through a complete neuropsychological evaluation, and despite the data obtained from such evaluation concluding that patients with PCA show specific needs and difficulties that require special attention and intervention.	

After performing a lumbar puncture, PCA and AD present with the same clinical findings (reduced level of β-amyloid 42 and increased levels of tau protein and phosphorylated tau in the cerebrospinal fluid). Likewise, in a postmortem study, patients of PCA present the same anatomopathological findings usually found in AD patients - the accumulation of neurofibrillary tangles and senile plaques. However, studies using PET, SPECT or MRI show that the neurodegeneration pattern in both diseases differs– while degeneration in AD begins in the temporomedial region, degeneration in PCA begins in the occipitoparietal cortex and affects the dorsal and ventral pathways of visual processing. These two different neurodegeneration patterns bring with them two differentiated clinical expressions, and, therefore, considering PCA as a pathological separate entity from AD could be a valid option.	
There are, indeed, very few reported cases of this disease. That does not mean that no other cases exist, but that they are not documented or published. This is why publishing single case studies is so important, as they are a valuable source of information for other clinicians, and therefore, the work presented by the authors is of interest. However, for the work to be published some corrections would be required.	
2.Methodology 2.1. The study has been carried out by an interdisciplinary team made up of an optometrist, a neurologist and an ophthalmologist, who met regularly to reach an agreement on the examination criteria. The authors should mention the specific tests performed, and the results obtained that led to the diagnosis of PCA.	
2.2. There is no mention whatsoever from the authors about what the patients' symptoms are, whether the symptoms of visual disturbances have had an insidious onset, whether the absence of an ophthalmologic pathology to explain the symptoms has been analysed, whether visual defects are disproportionate compared to other cognitive deficits, whether the course of the disease was progressive, whether tests were performed to prove the existence of unilateral or bilateral occipitoparietal or occipitotemporal atrophy, hypometabolism or hypoperfusion. Proving all this is essential to discard the existence of such a rare disease as PCA in neuropsychological tests at a later stage.	We have added some additional information relating to this. We have not sought to address every aspect of this comment as it was not the intention of this study to explore this level of detail of patient presentation / characterisation, but rather to explore qualitatively their experiences of the tests. These issues are of course likely to be important in any subsequent research.

2.3. The authors mention they have carried out a series of non-specified optometric tests. It would be advisable to know the visual exploration protocol performed on these patients, by both the optometrist and the ophthalmologist.	We have added this information.
2.4. The authors evaluate the visual tests as well or badly performed, but do not provide the values obtained from each test.	This study was not intended or designed to support any objective evaluation of the various tests - such evaluation clearly would have required larger numbers of participants, different vision assessment protocols (consistent test order etc.) and detailed recording of the values achieved, which could then be compared to a gold standard assessment potentially. We agree that such a study would be of interest, but felt that completing this exploratory pilot research would be useful in both demonstrating the utility of such a large projected, but also in providing some informative insights to the patients' and clinicians' experiences of the assessment process.
2.5. No references to fixation and saccades anomalies in the PCA are provided, despite the bibliographic resources available that support the alteration of these functions in this pathology. This could have affected the results of the tests performed.	Please see the comment above - this is a helpful observation in the design of future research.
3. The discussion about the results lacks a thorough reasoning and bibliographical supporting resources. Statements about the pathology are not referenced, and this is essential. The authors do not mention visual disturbances that are known to be experienced by the patients, such as prosopagnosia, apperceptive visual agnosia, or simultagnosia. There is no mention to which types of visuospatial deficits the patients have — optic ataxia or ocular apraxia. These are all vital signs when evaluating a patient's disability, whose importance is highlighted by the authors when stating that the prevalence of visual impairment was found to be more than 2.5 hours in residential settings.	We have revised the manuscript to add further refereces that we hope will address this point. We note however, that with the exception of the neurologist, the health professional participants were not engaged in delivering expert assessments of neurological assessments - this was rather the point of the study, since many people living with PCA encounter professionals who while highly experienced at assessing vision, may not have had the opportunity to encounter numbers of patients with visual impairment or difficulties that are neurological in origin.
4. Conclusions From the tests carried out to obtain the results of the current study and the explanations given by the authors, it cannot be concluded that 'a simple test which compared full and fragmented images or letters was agreed to be the test that provided the clearest evidence of PCA, or of symptoms of other cortical vision problems, as patients could identify the full image but nor the fragmented one.'	We have amended the draft to address this point - we have tried to make additionally clear that this conclusion emerged as the consensus of the clinician participants.

5. References The authors should do a more comprehensive literature review and discuss the results, as the current bibliography is insufficient and has little relation to visual recognition in PCA.	We have revised the literature review and references.
Reviewer: 1 LS This paper used 3 samples to carry out a pilot study about PCA and was trying to find how different tests are used to assess vision for people with PCA and whether a more effective test exists among them. With efforts of the researchers, some conclusions were drawn in the study. They found that simple and short tests were better and more effective than more subjective ones. Also, during the test, patient's vigour should be an important factor that might affect the result. The largest limit of the study is the small number of samples involved. In the future, researchers can recruit more age and gender matched individuals with different diseases, e.g., dementia, as well as control group for more comprehensive comparisons.	We appreciate these comments and believe that we have addressed them in our responses to previous reviewer comments, and that have been addressed within the revisions already discussed.

VERSION 2 – REVIEW

REVIEWER	Elena Salobrar-Garcia Universidad Complutense de Madrid, Spain
REVIEW RETURNED	17-Aug-2018

GENERAL COMMENTS	Authors improve the paper taking into account all the comments of the reviewers. In the paper there are a couple of grammar mistakes (The reviewer provided a marked copy with additional comments. Please contact the publisher for full details.)