

The Stages of Posterior Cortical Atrophy (PCA)

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Aims

When given a diagnosis of PCA, many people naturally ask what will happen next, how long will they be able continue with a particular activity, or when and what care will they need. Such questions are often met with the response 'we don't know' or 'it's different for each person'. In many ways these responses are reasonable, as the experience of PCA can indeed vary greatly from one person to the next. Symptoms may vary from individual to individual owing to a variety of factors including age, health and the disease underlying the PCA syndrome. Not everyone will experience the same symptoms or progress at the same rate. But whilst true, these responses are not terribly helpful.

This document attempts to provide a general framework for describing how abilities may change during the course of PCA. It is hoped that terms such as the 'stages' described may provide a common language which helps when discussing which treatments, therapies, support or decisions are appropriate.

The framework

This description of PCA is based on a seven-stage framework describing the progression of typical Alzheimer's disease (see http://www.alz.org/alzheimers_disease_stages_of_alzheimers.asp?type=alzFooter), and is derived from a system developed by Barry Reisberg, M.D., clinical director of the New York University School of Medicine's Silberstein Aging and Dementia Research Center. These original descriptions of the stages of typical AD are reproduced in an Appendix II at the end of the document to enable comparison between the two conditions. For some individuals, PCA may increasingly resemble typical AD as the condition progresses. For others, the changes experienced may remain partially distinct until the latest stages.

- Stage 1: No impairment (everything is normal)
- Stage 2: Very mild decline (something is wrong but it is not clear what; may include problems only recognized in hindsight)
- Stage 3: Mild decline (everyone accepts something is wrong; it is not wrong all the time but there are particular things that someone cannot do or problems they experience)
- Stage 4: Moderate decline (problems are now quite debilitating and help is required with a number of everyday activities)
- Stage 5: Moderately severe decline (problems are now seriously debilitating and help is required with most everyday activities)
- Stage 6: Severe decline (significant problems are experienced across a range of thinking and sensory abilities, and a more global dementia is evident)
- Stage 7: Very severe decline (many basic abilities are lost, and care needs resemble those of individuals with end-stage typical Alzheimer's disease)

Caveats

Before reading the descriptions of each stage, it is worth bearing in mind a few caveats:

- (a) As noted above, everybody is different. Rather than denying this difference, we will present the framework and then add commentaries from individuals with PCA and their carers to describe how well these stages describe their experience, or indeed the ways in which their experience has diverged from the course described below.
- (b) The definition of stages is inevitably arbitrary. The act of drawing lines between disease stages and placing them in a numerical order implies that progression through such stages will be gradual and linear. However some people will experience some symptoms in an order other than that described.
- (c) People may also move through some stages at a slower or faster rate. For this reason, the duration of each stage has not been estimated, but it is hoped that once a framework is established, it will make it easier for us to estimate the average speed at which the condition progresses.
- (d) Remember some people may be diagnosed earlier or later in the overall course of their PCA, so may not necessarily 'begin' their journey with label of PCA at Stage 1. For example, you might find that you have just been diagnosed but already experience many of the problems described in Stages 3 or 4.
- (e) It is also worth remembering that whilst these stages describe a progressive decline in particular skills, some people with PCA exhibit important coping mechanisms and/or resort to other more preserved skills to help themselves cope with aspects of their condition.
- (f) With regard to the language used to describe the stages (e.g. 'mild', 'moderate' and 'severe') it is important to recognize that perspectives change. For example, one's perception of what seemed 'severe' in the early days after a diagnosis might seem very different 8 or 10 years later in the illness. Without either belittling the challenges in the mild stages or scaring people by the prospect of what may be to come, it needs to be understood that one's perception of 'severity' may change.
- (g) It is important to note that often families and carers of people with PCA will notice sudden declines in abilities followed by a plateau in the symptom development rather than gradual changes in abilities.
- (h) Additionally, some symptoms are variable and not necessarily specific to PCA although they may be experienced fairly frequently by people with PCA. We have included these symptoms in the appendices (for example, certain people may experience weight loss or hallucinations).

The Stages of PCA

Stage 1: No impairment (normal function)

The person does not experience any visual problems, memory or other cognitive problems. An interview with a medical professional does not show any evidence of symptoms of vision loss or dementia.

Stage 2: Very mild cognitive decline (may include problems only recognized in hindsight)

The person experiences subtle difficulties with some complex visual tasks e.g.

- a loss of confidence in their driving ability owing to finding it harder to judge distances
- problems using a computer such as being slower to type or enter information
- difficulties with reading, writing, spelling and/or arithmetic
- difficulty recalling and writing down telephone numbers
- difficulty assembling novel devices/objects

The person may experience vertigo or other balance problems (although some aspects of these may develop later or not at all), they may experience difficulties finding the right words when speaking or odd visual sensations (e.g. occasional changes or washes of colour in the centre or periphery of their vision).

These problems may be intermittent, difficult to describe and not noticed by friends, family or co-workers. Insight into, but uncertainty as to the cause of these experiences may contribute to anxiety in some individuals. This stage may reflect the earliest signs of PCA, especially where eye tests reveal no indication of ophthalmological problems.

Stage 3: Mild cognitive decline (Early-stage Posterior Cortical Atrophy can be diagnosed in some, but not all, individuals with these symptoms)

Visual difficulties are acknowledged both by the person in question and their friends, family and co-workers, and the person is likely to decide to stop driving (although some people may have already stopped by this stage). Healthcare professionals familiar with young onset and atypical dementias may recognize a fundamental difficulty in perceiving what and where things are leading to characteristic problems including:

- Difficulty seeing things which are 'right under your nose' or only seeing part of something (e.g. not noticing food on the edge of a plate)
- General 'clumsiness' (in people who were not previously clumsy), such as knocking things over (inaccurate reaching out), putting a glass down sideways, etc.
- Difficulty finding things in a handbag, cupboard, etc.
- Difficulties dressing and performing other activities (e.g. DIY, cutting a piece of toast in half) that require spatial judgement (e.g. using stairs) and (e.g. left-right) coordination
- Inability to tell the time from a clock or watch (especially analogue)
- Getting 'lost' on the page when reading
- Problems with writing, spelling and dealing with numbers
- Paying for items with a note or letting vendors select coins for them (to avoid having to handle change)
- Mild memory problems for recent events, though less pronounced than visual problems – these could occur for different reasons, e.g. able to remember many events very well but difficulty for example learning new tasks
- Mild difficulty retrieving the correct word in conversation

These difficulties may be accompanied by anxiety; this may partly reflect awareness of the difficulties, but equally anxiety and feeling flustered can exacerbate the visual and other problems. People at this stage may also experience subtle sensory changes and disturbances (e.g. feeling cold).

Stage 4: Moderate cognitive decline (Mild or early-stage Posterior Cortical Atrophy)

At this point, individuals with PCA may require significant help with everyday activities such as cooking, dressing and using appliances. The person retains a clear sense of purpose regarding activities and plans but attempts to achieve these goals are frustrated by frequent, small visual problems and have significant implications for independence and the care/support required e.g.

- Inability to read labels on food packets
- Problems discriminating coins
- Inability to tell the time from an analogue or even digital watch

There may be significant contrasts between what people can and cannot do e.g.

- Dissociation in ability to walk, sit down and stand up (e.g. might be able to walk but not sit down or unable to stand up and walk).
- Being able to see some things but not others (e.g. following moving targets better than localizing static objects; can't read but can still follow or at least get pleasure from TV or the cinema)

There may be changes in the ability to navigate around unfamiliar and familiar environments: Someone with PCA might be aware that they are at home but have difficulty navigating from one room to the next, or locating the bathroom; some may find a loss in their mental map resulting in problems navigating even familiar environments, (e.g. the home) or problems locating rooms (especially in low light). This might result in getting lost within the home. Problems outside the house might also occur.

Severe visual and other problems may also result in:

- Inability to engage in a number of pastimes such as reading
- An inability to identify even very familiar faces, with recognition of identity dependent upon hearing someone's voice
- Resorting to eating with one's hands, or being fed, owing to difficulty manipulating cutlery and seeing the location of food on a plate

In addition, increasing word finding difficulties may mean that more time is required to express oneself, leading to reduced participation in group conversations.

Stage 5: Moderately severe cognitive decline (Moderate or mid-stage Posterior Cortical Atrophy)

Vision deteriorates further to the extent that the world is viewed as individual pieces of a puzzle or as if through a fractured mirror, with occasional small details noticed but difficulty appreciating whole scenes or the relationship between different objects. Individuals with PCA may now:

- Require help with most or all everyday tasks, including toileting and personal care owing to problems undoing zips/buttons, positioning the body relative to furniture and seeing and feeling the position of clothing, etc.
- Experience spatial problems not limited to vision, with spatial commands and actions (e.g. 'turn round', 'shuffle forwards') difficult to comprehend and execute
- Require support and/or supervision when walking, for example because of difficulty detecting the edge of pavements, paths and steps etc., and/or because of a tendency to shuffle, stoop or lean

At this stage, many individuals are registered as blind, meaning they require support in all visually guided activities. Episodic memory and word finding difficulties may also now be prominent, though many individuals retain considerable awareness of their situation. Some people may have problems with multiple senses, e.g. problems with:

- Sensing the relative position of parts of the body (as pronounced as not knowing if one is 'the right way up')
- Headache, increased sensitivity to pain and/or sensations in the scalp or other body parts, people with PCA may become more sensitive to the cold and may be prone to experiencing cold hands and feet
- Sense of hearing may change: people with PCA may become more sensitive to certain sounds.
- Feelings of imbalance or instability, particularly when walking
- Little jerky movements in the fingers, arms or other parts of the body

Stage 6: Severe cognitive decline (Moderately severe or mid-stage Posterior Cortical Atrophy)

At this stage, cognitive changes are more global in nature, with multiple aspects of perception, memory, language, attention and decision-making abilities affected. Vision remains the most pronounced impairment, but in most individuals, there are widespread impairments that would be recognised as dementia. Individuals may:

- Be 'functionally blind', requiring support in all visually-guided activities
- Show inconsistent recollection of recent events and plans
- Show halting, non-fluent speech with difficulty retrieving many words and the production of sound-based speech errors
- Experience changes in sleep patterns — sleeping during the day and becoming restless at night.
- Have increasingly frequent trouble controlling their bladder or bowels
- Become quiet and withdrawn, sometimes sitting in a stooped over manner and appearing disengaged from the environment unless addressed directly
- Experience behavior and personality changes, including obsessions, compulsions, suspiciousness and delusions (such as believing that their caregiver is an impostor).
- Have difficulty communicating, as expressing and understanding verbal and visual stimuli becomes increasingly difficult.
- Experience continued deterioration of sensory functions, e.g. partial or complete loss of response to touch

Stage 7: Very severe cognitive decline (Severe or late-stage Posterior Cortical Atrophy)

In the late stages, the problems experienced by people with PCA may resemble typical AD more closely than at any stage earlier in the disease process. In the final stage of this disease,

individuals lose the ability to respond to their environment, to carry on a conversation and, eventually, to control movement. They may still say words or phrases. There may be discrepancies e.g. inability to smile but continued ability to laugh. At this stage, individuals need help with much of their daily personal care, including eating or using the toilet. They may also lose the ability to smile, to sit without support and to hold their heads up. Reflexes become abnormal. Muscles grow rigid. Swallowing is impaired. Maintaining adequate nutrition, hydration and skin integrity can be challenging at this stage but problems (e.g. bedsores etc.) are not inevitable.

End of life

Although Alzheimer's disease and other degenerative diseases are life shortening illnesses, another condition or illness (such as pneumonia) may actually cause the person's death. Pneumonia is listed as the cause of death in up to two thirds of people with dementia. The person's ability to cope with infections and other physical problems will be impaired due to the progression of the disease. In some people no specific cause of death is found, other than Alzheimer's disease. Depending on the circumstances, 'Alzheimer's disease' or similar may be entered on the death certificate as the sole or main cause of death, or as a contributing factor.

Appendix I: Feedback from members of the PCA Support group

– “The above stage descriptions are apt but one of my wife's major symptoms, vivid hallucinations, does not feature strongly”

– “Overall, I think the above matches pretty well our experience of PCA so far, although for us, there is some overlap between the symptoms of each stage. The biggest difficulty with it is the inconsistencies in the symptoms of PCA with dad - for example, despite not being able to dress himself, there are still the occasional rare times when he can do it (or at least, most of it)...Also, I think it may be helpful to say that dad doesn't appear to decline gradually - more that abilities suddenly 'disappear' (in fact, overnight in the case of writing) and then he plateaus at that level for a time. I was expecting a constant, steady decline and was unprepared for the sudden nature of his loss of abilities when they happened”

– ‘[He] got very confused about dates and found his diary pretty confusing. And he would also get much more tired than 'normal' - as a result, we believed he was suffering from depression...dad also 'forgot' how to swim on a holiday - I now assume that it was more the result of the required coordinated nature of arms and legs, rather than him actually 'forgetting' how to do it. On that same holiday, he froze in airports when faced with shiny marble floors and took some convincing that he could walk on them. He reported puddles as holes at this point too.... He also reported odd visual disturbances to do with his hands and/or feet - for example, for a while he said that his hand was on 'backwards' or he didn't have all his toes/fingers etc. He also consistently reported being cold - even on a warm day (still does).’

– It's only the last stage I want to talk about. I really think there should be a stage 7A and stage 7B. Brian* uses his eyes more now than he did for the whole of last year. He appears to see things and people, sometimes saying 'Look' and pointing, but unable to say what he sees. Certainly reacts to people and to environment very well. Understands his situation. Still trying to lift his upper body. Holding his head well in the correct position until he gets tired, yet last year his head was down nearly the whole time! Never saw him with his eyes open last year. Still able to grasp well with his hands, though less with his left. Still produces short whole sentences, will answer direct questions about 50% of the time. Both smiles and laughs appropriately. Recognizes different people, for example knows very well when the Home Manager is in the room. Swallows well. Shows clear liking or disliking for particular foods. Has not apparently lost any sensitivity to touch. Has very obvious recall of recent events. Nevertheless, he's unable to do anything for himself. I also could have added that Brian is taken three times a week to a local day centre, comes home once a week, and has other odd outings, all by wheelchair taxi. He has a physio twice a week, has had a dietician, has a variety of equipment, and a private carer as well as me. This probably all has an effect. His spoken vocab has been increasing recently. I maybe also should have added that our biggest current problem is teeth grinding.

*name changed

– “[The stages fit] Quite well - have indicated any discrepancies. You have not made mention of weight and diet change which were quite noticeable with Alice* (loss of weight

and change in diet and portions). Also, I wonder if it would be worth mentioning certain physical symptoms which could be related - Alice had constant stomach and bowel problems in the earlier stages; which despite many GP and hospital visits remained unresolved. We feel these must have been caused by anxiety. Then later on, she had many UTIs. As Alice was older than the typical PCA-er - in her retired years when symptoms became noticeable: I feel diagnosis took longer. This was partly her own fault for 'hiding' symptoms from us, but also due to her GP and other professionals stating that her problems were 'normal age-related degeneration' and 'a case of conversion disorder (hysterical blindness)'. There were some symptoms that she never showed such as jerking limbs or even (as far as we were aware) short-term memory loss. And thankfully, although clearly highly anxious, she never showed any personality change. “

*name changed

Appendix II. The original 'Seven stages of Alzheimer's disease' as described for people with a more typical presentation of AD

Stage 1: No impairment (normal function)

The person does not experience any memory problems. An interview with a medical professional does not show any evidence of symptoms of dementia.

Stage 2: Very mild cognitive decline

The person may feel as if he or she is having memory lapses — forgetting familiar words or the location of everyday objects. But no symptoms of dementia can be detected during a medical examination or by friends, family or co-workers. This stage may reflect normal age-related changes or the earliest signs of Alzheimer's disease.

Stage 3: Mild cognitive decline (Early-stage Alzheimer's can be diagnosed in some, but not all, individuals with these symptoms)

Friends, family or co-workers begin to notice difficulties. During a detailed medical interview, doctors may be able to detect problems in memory or concentration. Common stage 3 difficulties include:

- Noticeable problems coming up with the right word or name
- Trouble remembering names when introduced to new people
- Having noticeably greater difficulty performing tasks in social or work settings
- Forgetting material that one has just read
- Losing or misplacing a valuable object
- Increasing trouble with planning or organizing

Stage 4: Moderate cognitive decline (Mild or early-stage Alzheimer's disease)

At this point, a careful medical interview should be able to detect clear-cut symptoms in several areas:

- Forgetfulness of recent events
- Impaired ability to perform challenging mental arithmetic — for example, counting backward from 100 by 7s
- Greater difficulty performing complex tasks, such as planning dinner for guests, paying bills or managing finances
- Forgetfulness about one's own personal history
- Becoming moody or withdrawn, especially in socially or mentally challenging situations

Stage 5: Moderately severe cognitive decline (Moderate or mid-stage Alzheimer's disease)

Gaps in memory and thinking are noticeable, and individuals begin to need help with day-to-day activities. At this stage, those with Alzheimer's disease may:

- Be unable to recall their own address or telephone number or the high school or college from which they graduated
- Become confused about where they are or what day it is
- Have trouble with less challenging mental arithmetic; such as counting backward from 40 by subtracting 4s or from 20 by 2s

- Need help choosing proper clothing for the season or the occasion
- Still remember significant details about themselves and their family
- Still require no assistance with eating or using the toilet

Stage 6: Severe cognitive decline (Moderately severe or mid-stage Alzheimer's disease)

Memory continues to worsen, personality changes may take place and individuals need extensive help with daily activities. At this stage, individuals may:

- Lose awareness of recent experiences as well as of their surroundings
- Remember their own name but have difficulty with their personal history
- Distinguish familiar and unfamiliar faces but have trouble remembering the name of a spouse or caregiver
- Need help dressing properly and may, without supervision, make mistakes such as putting pajamas over daytime clothes or shoes on the wrong feet
- Experience major changes in sleep patterns — sleeping during the day and becoming restless at night
- Need help handling details of toileting (for example, flushing the toilet, wiping or disposing of tissue properly)
- Have increasingly frequent trouble controlling their bladder or bowels
- Experience major personality and behavioral changes, including suspiciousness and delusions (such as believing that their caregiver is an impostor) or compulsive, repetitive behavior like hand-wringing or tissue shredding. The person may also repetitively articulate certain words or sounds.
- Tend to wander or become lost

Stage 7: Very severe cognitive decline (Severe or late-stage Alzheimer's disease)

In the final stage of this disease, individuals lose the ability to respond to their environment, to carry on a conversation and, eventually, to control movement. They may still say words or phrases. At this stage, individuals need help with much of their daily personal care, including eating or using the toilet. They may also lose the ability to smile, to sit without support and to hold their heads up. Reflexes become abnormal. Muscles grow rigid. Swallowing is impaired. Maintaining adequate nutrition, hydration and skin integrity can be an issue at this stage.

End of life

Although Alzheimer's disease and other degenerative diseases are life shortening illnesses, another condition or illness [such as pneumonia] may actually cause the person's death. Pneumonia is listed as the cause of death in up to two thirds of people with dementia. The person's ability to cope with infections and other physical problems will be impaired due to the progression of the disease. In some people no specific cause of death is found, other than Alzheimer's disease. Depending on the circumstances, 'Alzheimer's disease' or similar may be entered on the death certificate as the sole or main cause of death, or as a contributing factor.