

Posterior Cortical Atrophy

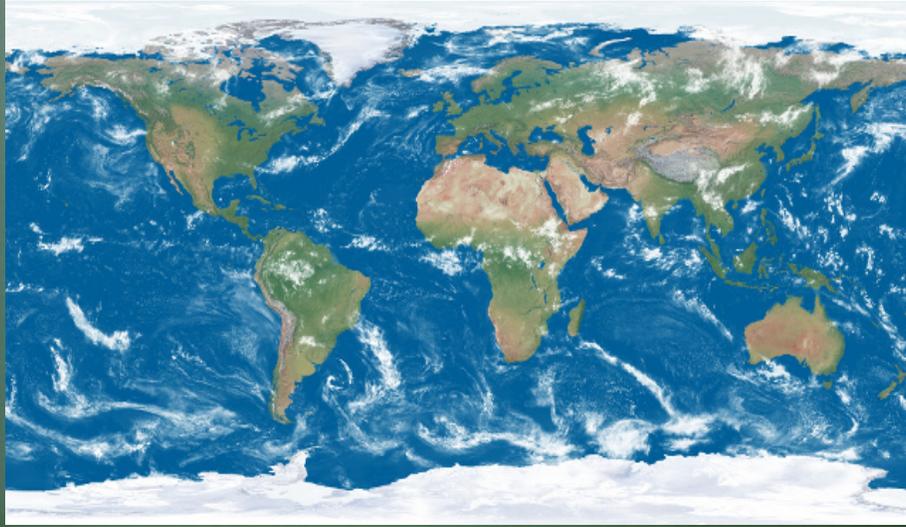


Posterior Cortical Atrophy

What is PCA?



Epidemiology - how rare and
where is it?



The prevalence and incidence of PCA are currently unknown; obtaining these data will depend on the adoption of consistent diagnostic criteria.

Neuropsych and clinical features – what are the symptoms?

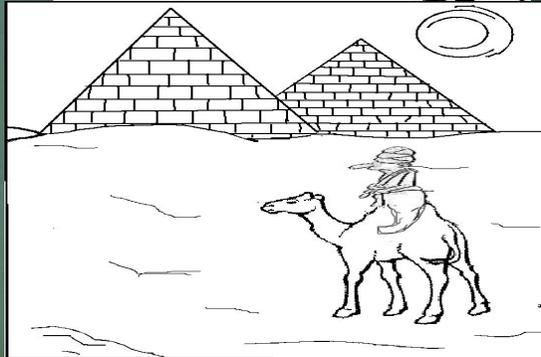
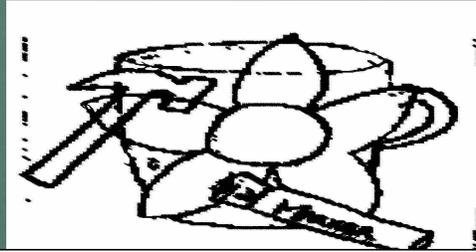
- Visuospatial and visuo-perceptual impairments
- Alexia

~~WRITTEN~~

~~WORD~~

Symptoms: Bálint's syndrome

- Simultanagnosia
 - dorsal
- oculomotor apraxia
- optic ataxia
- environmental agnosia



Oculomotor apraxia (OMA), also known as Cogan's Ocular Motor Apraxia or Saccadic Initiation Failure (SIF) is the absence or defect of controlled, voluntary, and purposeful eye movement.

Simultanagnosia (or simultagnosia) is a rare neurological disorder characterized by the inability of an individual to perceive more than a single object at a time. Dorsal (cannot see two things at once), top, ventral (cannot identify two things at once), bottom optic ataxia (lack of coordination between visual inputs and hand movements, resulting in inability to reach and grab objects)

Environmental agnosia - The syndrome is characterized by an inability to recognize familiar surroundings in spite of relatively intact verbal memory, cognition, and perception.

Symptoms: Gerstmann's
syndrome

- acalculia
- agraphia
- finger agnosia
- left-right disorientation

$$3x + 4 = 22$$

$$x = ?$$



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Acalculia: Loss of the ability to perform simple arithmetic calculations, typically resulting from disease or injury of the parietal lobe of the brain.

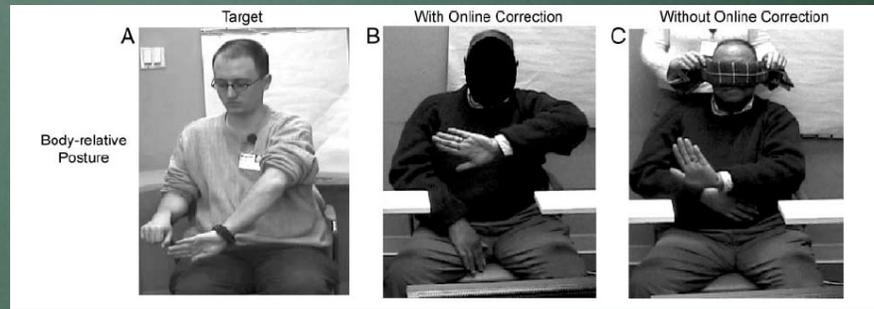
Agraphia: a loss of the ability to write or to express thoughts in writing because of a brain lesion.

Finger agnosia, first defined in 1924 by Josef Gerstmann, is the loss in the ability “to distinguish, name, or recognize the fingers”, not only with the patient’s own fingers, but also the fingers of others, and drawing and other representations of fingers.

left right disorientation
an inability to distinguish the left from the right;

Symptoms

- Deficits in working memory
- Limb apraxia



SOURCE: haptics.grasp.upenn.edu

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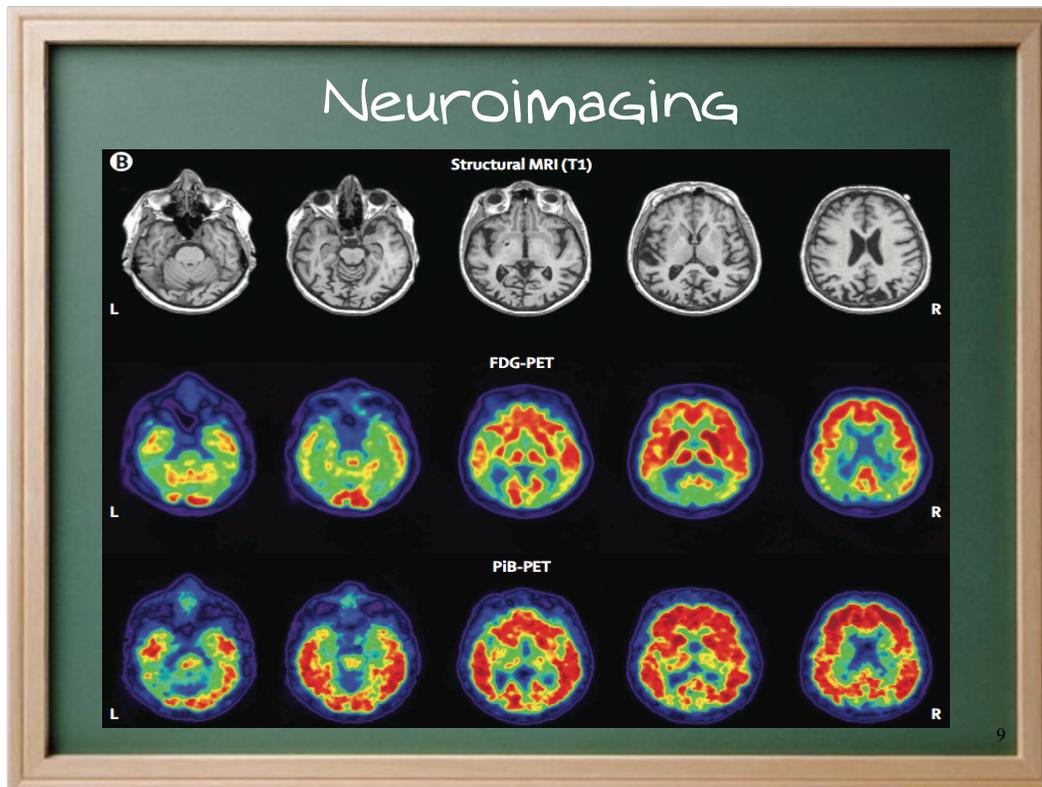
Working memory is the system that actively holds multiple pieces of transitory information in the mind, where they can be manipulated.

Limb apraxia is a dysfunction in a learned movement of a limb.

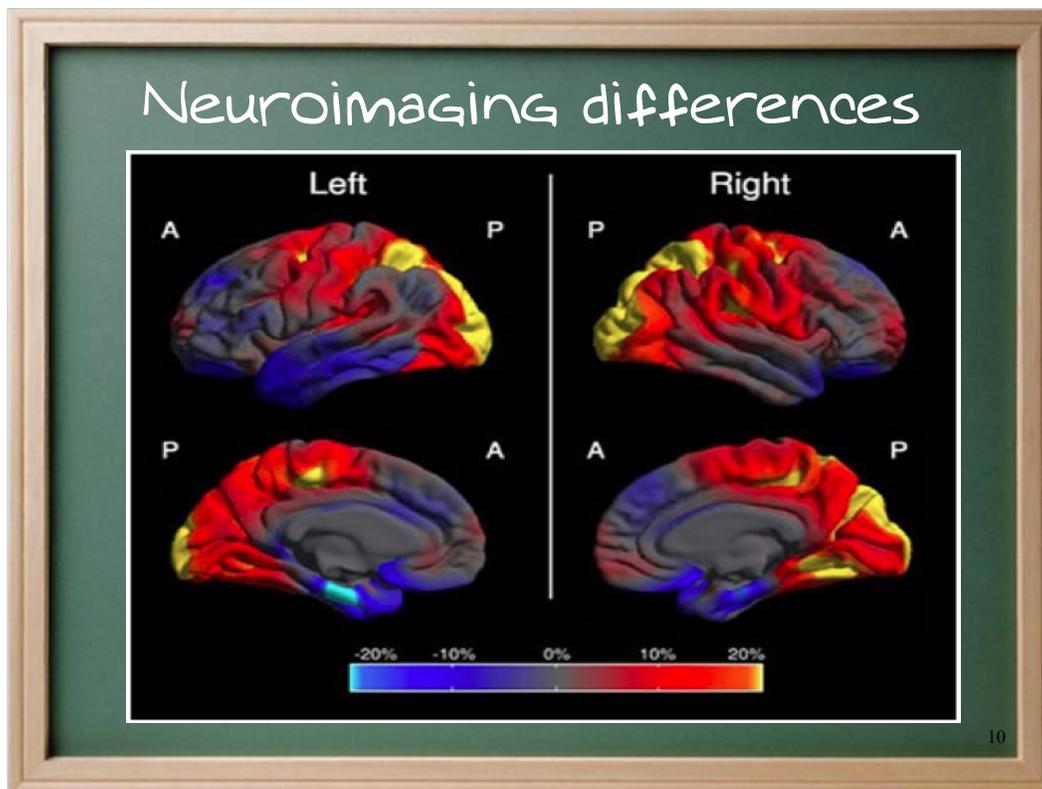
Possible Later Symptoms

- Typically preserved better in early PCA than in typical Alzheimer's Disease
 - anterograde memory deficit
 - executive function deficit
 - linguistic skills deficit

Findings of longitudinal studies indicate that these, which are sometimes preserved strikingly in earlier stages of disease, gradually deteriorate in some patients with PCA as they progress to a more global dementia state.



Brain MRI showed striking atrophy in bilateral parietal, posterior temporal, and lateral occipital cortex (figure 2B [upper row]), and fluorodeoxyglucose (FDG)-PET (figure 2B [middle row]) showed hypometabolism in the same regions, left worse than right. Frontal cortex, medial temporal cortex, and hippocampus were spared. Pittsburgh compound (PiB)-PET showed diffuse cortical uptake throughout posterior and anterior cortical regions alike (figure 2B [lower row]), consistent with underlying amyloid- β plaques.



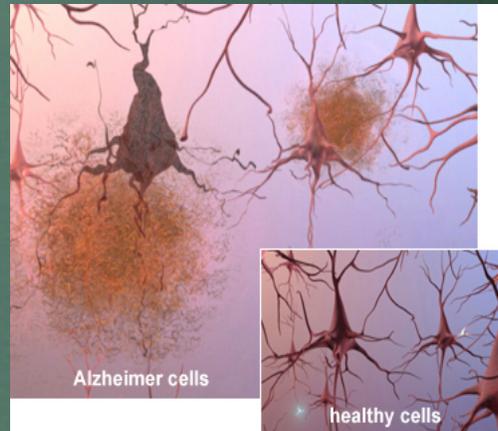
This image shows in which regions of the brain the cortex is abnormally thin in people with PCA compared to people with AD. The color scale represents the magnitude of the difference in cortical thickness; left and right refer to the brain's hemispheres. Yellow and red represent more cortical thinning in PCA (occipital and parietal cortex); blue represents more cortical thinning in AD (mediotemporal, entorhinal, frontal region). Summary image from 48 PCA and 30 AD cases, derived from Figure 1 in Lehmann et al., 2011.

Genetics

- PCA is strongly associated with Alzheimer's Disease (AD) genetic links
- No specific PCA genetic subtype of AD has been discovered

Pathology

- Alzheimer's Disease is most common cause
 - Plaques
 - tangles



SOURCE: http://www.alz.org/braintour/plaques_tangles.asp

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- a) Plaques, abnormal clusters of protein fragments, build up between nerve cells. - amyloid protein snippets normally vacuumed in healthy brain
- b) Dead and dying nerve cells contain tangles, which are made up of twisted strands of another protein – in abnormal tau protein, normal microtubule structure collapses
 - i. Plaques and tangles are more likely in the occipital lobe in PCA than canonical Alzheimer's disease, but elsewhere in the brain distribution is same

Diagnostic Criteria

- Syndrome
- Proposed symptoms:
 - Insidious onset, gradual progression
 - Visual deficits w/o ocular disease
 - Other symptoms listed earlier
- Syndrome diagnosis considered incomplete, need to know pathology

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How do we determine if someone is suffering from it?

Insidious onset is a disease that comes on slowly, without obvious symptom at first so the person is not aware of its development.

Management

- Alzheimer's drugs (acetylcholinesterase inhibitors) unevaluated, likely effective
- Antidepressant and Parkinsonism drugs recommended for symptoms
- Support groups, compensations for visual difficulties critical
- Keeping Brain active helpful

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How can it be treated?



"Sometimes he cannot see the cup that is in front of him. Clumsiness is another of the symptoms. When he tremblingly puts his glass of juice down on a glass-topped table in his hotel suite, it makes a crash, as if he misjudged the distance, but then glass-topped tables are tricky. I might easily have done the same. You don't know with Pratchett, and often nor does he, how much to attribute to his disease and how much is natural ageing. So far, the cognitive parts of his mind seem untouched."

Advocacy Group

- Founded in 2012
- PCA is a clinical syndrome
- "With a little help they can do almost everything; without help they can't do anything."

Traditional Alzheimer's Groups

Inappropriate

- Younger, diff. challenges
- Activities offered (puzzles, large-font books) are visual and actively unhelpful for PCA sufferers



SOURCE: Blue Valley Lutheran Homes

Further Information

- http://www.alz.org/alzheimers_disease_4719.asp
- <http://www.alz.org/dementia/posterior-cortical-atrophy.asp>
- Shining a light on posterior cortical atrophy., Crutch SJ, Schott JM, Rabinovici GD, Boeve BF, Cappa SF, Dickerson BC, Dubois B, Graff-Radford NR, Krolak-Salmon P, Lehmann M, Mendez MF, Pijnenburg Y, Ryan NS, Scheltens P, Shakespeare T, Tang-Wai DF, van der Flier WM, Bain L, Carrillo MC, Fox NC.
Alzheimers Dement. 2012 Dec 28. doi:pii: S1552-5260(12)02522-8. 10.1016/j.jalz.2012.11.004. [Epub ahead of print]
- Commentary on "Shining a light on posterior cortical atrophy", Jellinger KA.
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- Posterior cortical atrophy., Crutch SJ, Lehmann M, Schott JM, Rabinovici GD, Rossor MN, Fox NC.
Lancet Neurol. 2012 Feb;11(2):170-8. doi: 10.1016/S1474-4422(11)70289-7. Review.

Further Information, Brains

- <http://www.brainfacts.org>
- <http://blog.arichneuraltapestry.com>