"Beyond the EYE - behavioral and cortical assessment in Posterior Cortical Atrophy (PCA)"

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Background: Posterior cortical atrophy (PCA) is a neurodegenerative syndrome characterized by a progressive, dramatic and relatively selective decline in higher visual processing. Age of onset tends to be young, typically 50-65 years old, and the underlying pathology is associated most commonly with Alzheimer disease. Even though the syndrome is being recognized for more than two decades, PCA is relatively neglected by clinicians and researchers, and the patients are often referred to recurrent ophthalmic evaluation and face considerable delays in diagnosis

**Objective: To identify the behavioral signature of PCA patients**

Methods: Six patients with PCA and Four age matched controls underwent comprehensive set of visual PCA and neuropsychological tests. Visual tests included- visual acuity, stereopsis, color and ocular-motor evaluation. High order visual evaluation included- visual motor skills (pointing), motion coherence, global perception, pop-out, and visual perception tests composed of spatial relations (SR), form consistency (FC), visual memory (VM), visual closure (VC), figure ground (FG) and visual discrimination (VD).

Three patients underwent fMRI scan, addressing the neuronal substrate of the visual functions. An additional PET scan
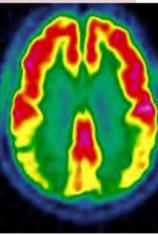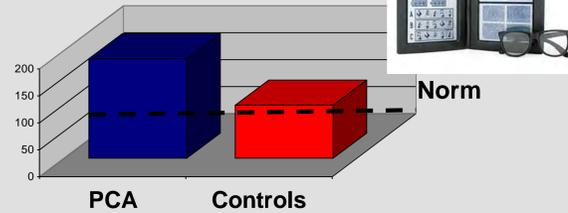
Results: low order visual and oculomotor skills

Developmental Eye Movement Test
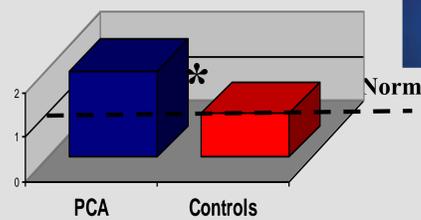
Ratio > 1 stands for impaired pursuit

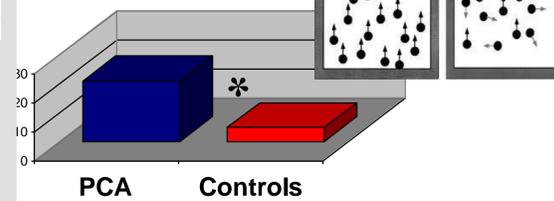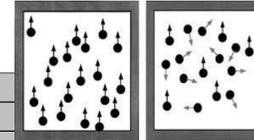**Stereopsis perception**

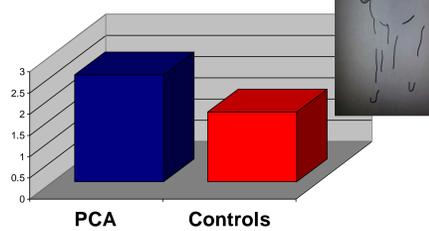Reduced stereopsis perception was evident in some patients

Results: High order Visual and visuo-spatial skills: A deficit in Global perception?
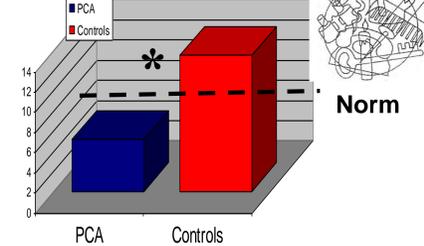
Motion Coherence

Impaired ability to perceive the global direction of an array of dots
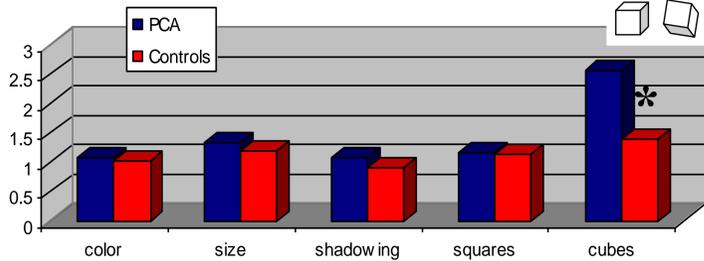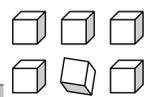
Visual closure (Golen)

Reduced ability to perceive visual images based on image's components

Figure ground-Tangled I

Impaired ability to differentiate between figure and ground

Pop out

Pop-out (the ability to immediately see the exceptional image inside an array of images) was impaired only for images composed of subjective contours ("Kanizsa" squares). Global perception is needed to perceive these kinds of stimuli

Visual perception (NMVPT)

Ratio correct
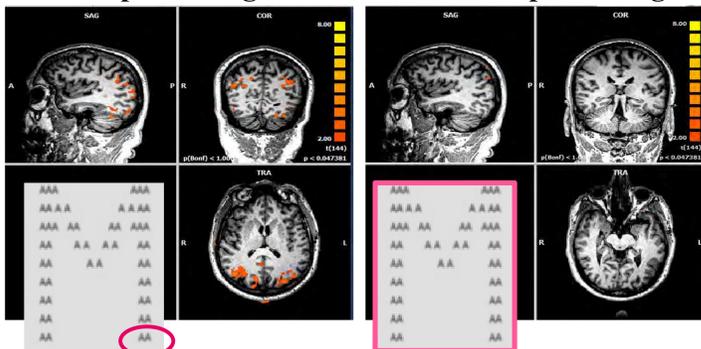
SR   FC   VM   VC   VD   FG
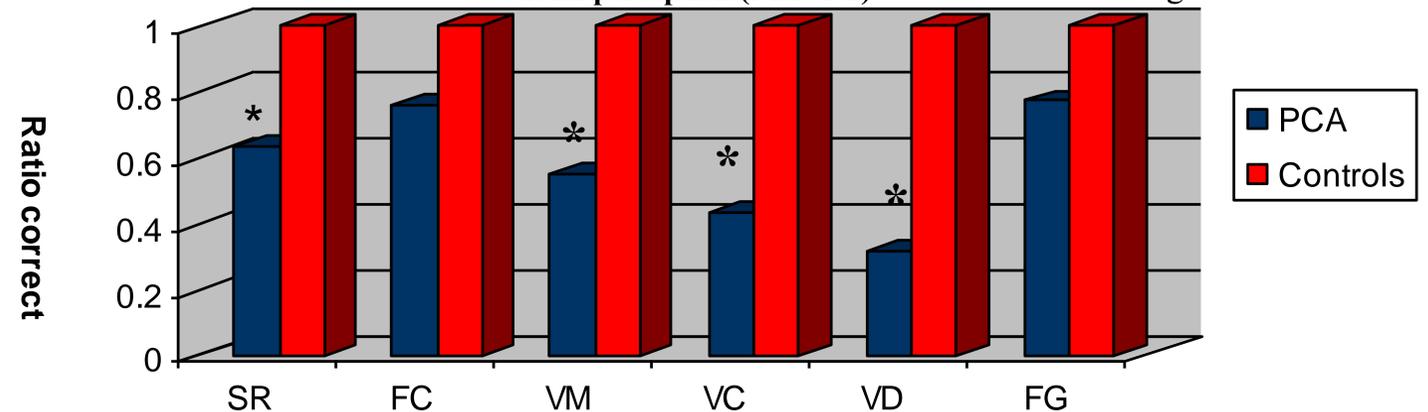
Impaired spatial relationship discrimination (SR), visual memory (VM), the ability to perceive an object based on its visual components (VC), and impaired discrimination of fine visual details (VD) were evident

Visual cortical activation (fMRI)

Local processing   Global processing

Reduced fMRI activity was found when subjects were asked to refer to the Global image (the "M") as compared to refering to its components (the small A's, Navon letters).

Conclusions: Developmental Eye movement test (DEM), Non movement Visual Perception Test (NMVPT), Motion coherence, and figure ground testing with the tangled figure were impaired in PCA patients. These tests as well as the fMRI activation pattern suggest a specific deficit in global perception in this patients population.

Greater awareness of the syndrome is needed to improve diagnostic accuracy, clinical management and design of research studies.