

Case Report

# Statokinetic Dissociation (Riddoch Phenomenon) in a Patient with Homonymous Hemianopsia as the First Sign of Posterior Cortical Atrophy

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## Keywords

Homonymous hemianopsia · Statokinetic dissociation · Riddoch phenomenon · Posterior cortical atrophy

## Abstract

We report a 60-year-old woman with posterior cortical atrophy (PCA) who presented with left homonymous hemianopsia persisting for 5 years; the patient's condition was observed using static, but not kinetic, perimetry. This statokinetic dissociation of hemianopsia, which is often called Riddoch syndrome, might have been caused by a dysfunction of the right primary visual and visual association cortices, representing a functional imbalance within a disturbed visual cortex. In patients with PCA and visual field defects, both static and kinetic perimetry may be useful for understanding the extent of degeneration in the visual cortex, in addition to examinations of unilateral neglect.

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Statokinetic dissociation (SKD), which is often called Riddoch phenomenon or Riddoch syndrome, is the ability to perceive visual motion consciously in a blind visual field [1] and has been observed in individuals with lesions in the anterior visual pathways [2] or the occipital lobe [3]. Although a visual field defect, especially hemianopsia or quadrantanopsia, is a feature of posterior cortical atrophy (PCA) [4] and may progress for several years without additional signs [5], SKD has rarely been mentioned in the literature. Here, we report an individual with PCA presenting with left homonymous hemianopsia observed using static, but not kinetic, perimetry, suggesting that the mechanism of SKD in PCA is worth discussing.

A 60-year-old right-handed housewife was referred to our hospital because of left homonymous hemianopsia without visual complaints after undergoing a 2-year annual check-up for glaucoma. Other than hemianopsia (Fig. 1a) and a visual acuity of 20/500 in both eyes, her intraocular pressure, funduscopy and Goldmann field test findings were almost normal (Fig. 1b). Her past and family history, a physical and neurological examination, and brain magnetic resonance imaging (MRI) were unremarkable. Subsequently, regular ophthalmic examination revealed that her visual field defects had progressed to involve the right lower quadrant on the Humphrey test. Although sequential optical coherence tomography examinations revealed a diffuse thinning of the retinal ganglion cell complex layer, especially in the peripheral areas, the areas of thinning did not correspond to the visual field defect on the Humphrey test.

Five years after our initial examination, although her Goldmann test findings continued to be normal, the patient had gradually begun to exhibit difficulty understanding test instructions. She also became forgetful, despite her almost normal daily activity. At this time, her neurological examination was normal except for a mild difficulty in grasping objects on her left side. She scored 28/100 on the Addenbrooke's Cognitive Examination-Revised (attention and orientation, 7/18; memory, 5/26; language, 13/26; fluency, 1/14; and visuospatial, 2/16). She also exhibited difficulty recognizing pictures and characters. Her drawing and line bisection revealed mild left hemispatial neglect. Color and face recognition was moderately disturbed. She had not experienced aphasia, apraxia, or visual hallucinations. A brain MRI examination revealed right dominant bilateral occipito-parieto-temporal atrophy (Fig. 1c). The patient next underwent a brain single photon emission computed tomography (SPECT) examination with Tc-99m ethyl cysteinate dimer. Using the easy z-score imaging system (eZIS; FUJIFILM RI Pharma, Tokyo, Japan) [6], the SPECT images were standardized using statistical parametric mapping and compared with those for age-matched controls included in the eZIS program. The z-score maps of our patient showed right dominant hemispheric hypoperfusion, with severe hypoperfusion observed in the occipito-parietal cortex and moderate hypoperfusion observed in the primary visual cortex (V1) and superior and middle temporal area (V5/hMT+) (Fig. 1d).

The concept of PCA was initially proposed by Benson et al. [7] as a unique type of dementia. PCA presents with progressive cortical visual dysfunction with a relative sparing of memory and exhibits a mainly Alzheimer disease neuropathology [4]. Based on the proposed diagnostic criteria [4], we diagnosed the present patient as having PCA.

Visual field defects in PCA have been reported to be less dense and less congruent, compared with defects in cases with stroke or trauma [8]. The visual field defect is thought to result from the degeneration of primary visual and visual association cortices [4, 9]. Since patchy neuronal dysfunction in the cortices of patients with PCA does not completely eliminate sensitivity to visual stimuli, kinetic perimetry may not be capable of revealing abnormalities until the defects become absolute [10]. Therefore, visual field tests using thresholds are more sensitive for detecting early defects in PCA [10], possibly accounting for the obser-

vation of SKD in the present patient with PCA. Findings similar to SKD have been reported in other patients with PCA [11, 12]. To our knowledge, however, such extraordinary prolongation of SKD as the only symptom of PCA for 5 years is quite atypical and has not been reported previously.

The unusual features of visual field defects in PCA may result from damage to the optic radiations and/or extrastriate cortex, rather than the V1 area [8]. Based on a multi-modal MRI analysis, Millington et al. [8] reported that the lateralized visual field defects observed in PCA were correlated with microstructural changes in optic radiations and the atrophy of extrastriate visual areas (V2 and V5/hMT+) in the hemisphere contralateral to the visual field defects. On the other hand, the mechanism of SKD arising from occipital lobe disease is assumed to involve visual inputs reaching the motion processing cortex (V5/hMT+) without passing through the V1 area, leading to a conscious awareness of motion within a blind field [1]. Although in the present case, the presence of right occipito-parietal hypoperfusion including the V1 and extrastriate cortex, as demonstrated using SPECT, may explain the homonymous hemianopsia, the hypoperfusion of the right V5/hMT+ area seems to contradict the presence of SKD in our patient. However, while unilateral V5/hMT+ or parieto-occipital lesions can reportedly cause akinetopsia (motion blindness), such phenomena rarely persist, probably because of the contribution of several cortical areas to global motion processing [13]. Therefore, compensation for V5/hMT+ dysfunction may be easier than compensation for V1 dysfunction, at least until the degeneration of all the motion processing areas, and this difference in the capacity for functional compensation for neuronal degeneration among visual cortical areas might have been responsible for the prolonged period of SKD in our patient.

Meanwhile, in daily clinical settings, the kinetic perimetry test is usually performed at a slower pace and is less affected by neglect [14]. Accordingly, unilateral neglect may account for SKD. Unfortunately, whether our case initially exhibited unilateral neglect was unclear because of the lack of neglect tests performed at the time of onset. However, since the visual field defects gradually spread to the contralateral field, it seems unlikely that spatial neglect was a major cause of the SKD in our patient. In addition, the areas of hypoperfusion in our patient did not involve the prefrontal region, which is presumed to be associated with hemineglect in PCA [15].

PCA can be another cause of SKD in occipital lobe disease. Clinically, this unique phenomenon may be the only initial sign, without any other symptoms, and may represent a functional imbalance among the degeneration of various visual processing areas. In patients with suspected PCA with visual field defects, both static and kinetic perimetry, in addition to neglect tests, may be useful for understanding the extent of degeneration in the visual cortex.

### Statement of Ethics

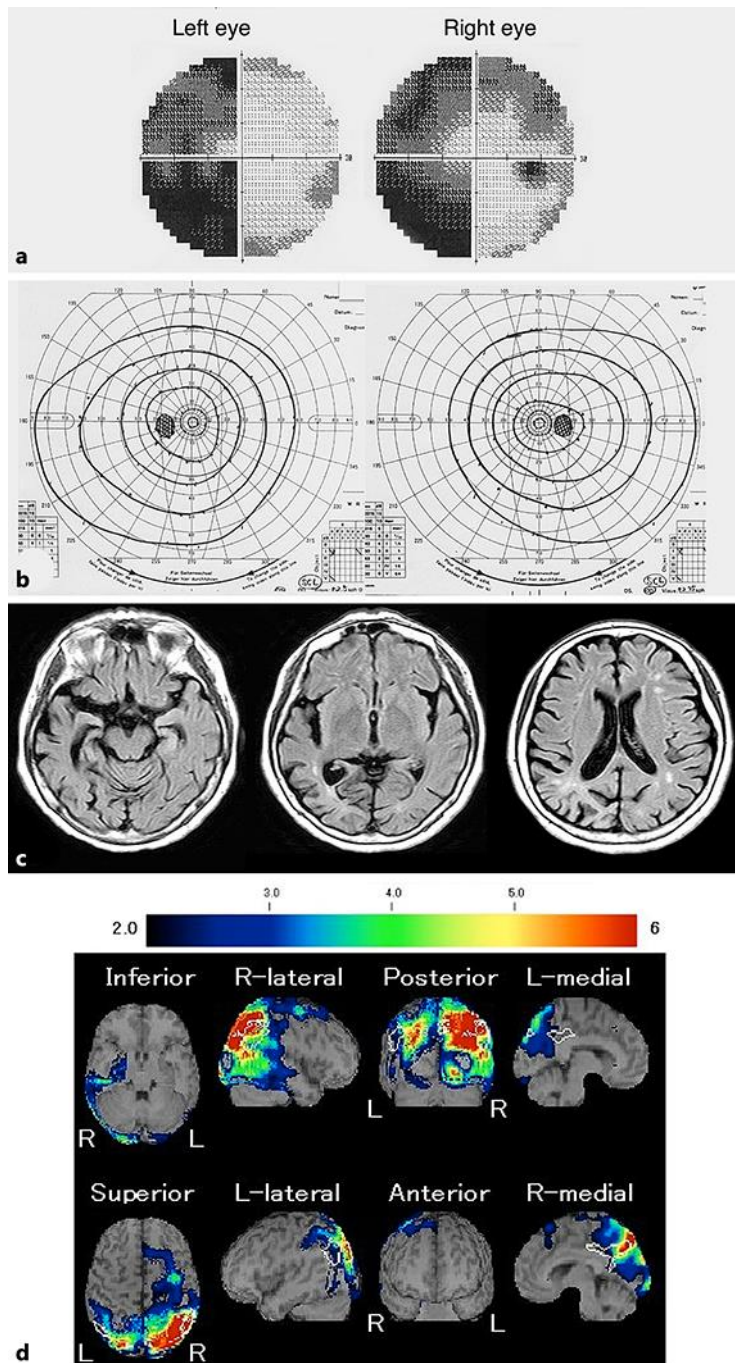
Written informed consent was obtained from the patient for the material to be submitted to the journal.

### Disclosure Statement

The authors declare that they have no conflicts of interest.

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**Fig. 1.** Humphrey visual field tests showed incongruent left hemianopia (a) with normal Goldmann visual fields (b) at the initial examination. After 5 years, axial MR images showed right dominant bilateral occipito-parieto-temporal atrophy (c). Using the easy z-score imaging system (see text), SPECT scans revealed right dominant hemispheric hypoperfusion, with severe hypoperfusion in the occipito-parietal cortex (d).