

protein synthesis are considered to cause selective vulnerability to hypoglycemia.<sup>16–19</sup> Changes in the basal ganglia observed on DWI of patients with hypoglycemic coma predict a poor outcome except for one report.<sup>2,3</sup> In the presented case, there was no signal change in the motor and sensory cortex on DWI. This allowed recovery of extremity movements. The motor, sensory and visual centers in the cortex are phylogenetically older than the association cortex,<sup>20</sup> which are supported by a lower energy in animals compared to humans.<sup>21</sup> This may explain the differential vulnerability to hypoglycemia, even in the human cortex.

#### 4. Conclusion

In profound hypoglycemic encephalopathy, vulnerability to hypoglycemia varies in different regions of the cortex. The DWI sequence of MRI can distinguish such areas of vulnerability and thus may be useful for predicting functional outcome.

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## Inconspicuous visual field defect in anterior choroidal artery territory infarction

Hsin-Hung Chen <sup>a</sup>, Chia-Yuen Chen <sup>b</sup>, Chi-Tzong Hong <sup>a,c,\*</sup>

<sup>a</sup> Department of Neurology, Taipei Medical University – Municipal Wan Fang Hospital, No. 111, Hsing-Long Road, Section 3, Taipei 116, Taiwan

<sup>b</sup> Department of Radiology, Taipei Medical University – Municipal Wan Fang Hospital, Taipei, Taiwan

<sup>c</sup> Brain Diseases and Aging Research Center, Taipei Medical University – Municipal Wan Fang Hospital, Taipei, Taiwan

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\* Corresponding author. Tel.: +886 2 29307930; fax: +886 2 29335221.  
E-mail address: kenhong88@yahoo.com.tw (C.-T. Hong).

## Abstract

We describe a patient with a right anterior choroidal artery territory infarction and an inconspicuous left visual defect. The anterior choroidal artery is a unique artery of the cerebral circulation. The occlusion of this artery can result in dysfunction of motor, sensory, and visual systems with only rare involvement of higher cortical function. Among symptoms reported, visual abnormalities are the most variable and the least common. However, the visual field abnormality may be overlooked and the incidence underestimated since some patients may not be aware of the problem until uncovered by formal visual field testing.

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## 1. Introduction

The anterior choroidal artery (AChA) is a unique artery of the cerebral circulation. The occlusion of this artery can result in dysfunction of motor, sensory, and visual systems.<sup>1,2</sup> Hence, hemiparesis, hemisensory impairment, and hemianopsia are referred to as the triad of AChA syndrome.<sup>1</sup> The complete form of this syndrome is not frequent, and one or more of the three cardinal symptoms may be missing.<sup>1,2</sup> Disturbances of higher cortical modalities are sometimes detected but they are usually transient.<sup>1,2</sup> A defect of the visual field, which is the most variable feature of the triad of AChA syndrome, occurs less frequently than other features of this syndrome.<sup>1,2</sup> Pure visual involvement is a very rare condition and only two cases have been reported.<sup>3</sup> Here, we describe a patient with a right anterior choroidal artery territory infarction and an inconspicuous left visual defect as the only persisting clinical manifestation.

## 2. Case report

A 74-year-old right-handed man noted an unsteady gait on getting up from his cabin seat after a short air trip. He also noted a painful sensation over the right periorbital area and scalp and slight slurring of his speech. He was transported immediately to the emergency room of a nearby hospital. A brain computerized tomographic (CT) scan was normal. Six hours later all symptoms disappeared although no specific treatment had been given. However, similar symptoms returned on waking the next morning. In addition, he felt sleepy and nauseated. He returned to the same hospital. A second brain CT scan was negative. Four hours later, he felt he had recovered completely. Throughout the course of these events no blurred vision had been reported or observed. Over the following 2 days the patient noted a protracted sense of heaviness on his head, and also felt nauseated. He was therefore referred to our neurology clinic.

The patient had no past history of stroke, peripheral vascular disease, diabetes, hypertension, arrhythmia, or heart disease. He had not smoked over the last 20 years and was not currently taking any medication. His family history was unremarkable.

On admission, blood pressure was 128/88 mmHg; temperature, 36.5 °C; heart rate, 70/min; respiration, 18/min.

No carotid bruits were audible, and examination of the heart and lungs was unremarkable, as was the rest of the general physical examination.

At the time of admission, neurological examination revealed normal orientation to person, time, and place. Repetition, comprehension, and use of language were normal. On confrontation test, a partial left homonymous hemianopsia was found in the absence of any subjective visual symptom. The functions of other cranial nerves were intact. Motor and sensory systems were normal. Cerebellar function was unaffected.

Laboratory studies revealed normal lipid profile, prothrombin time, and activated partial thromboplastin time. Erythrocyte sedimentation rate and C-reactive protein were not elevated. Perimetry conducted 3 days later disclosed a left incongruent homonymous hemianopsia more affecting the lower quadrant with central sparing (Fig. 1). Electrocardiography revealed normal sinus rhythm. The extracranial carotid duplex showed 99% stenosis of the right internal carotid artery. Magnetic resonance angiography of the brain revealed decrease of flow signal in the right internal carotid artery. Magnetic resonance images of the brain on T2-weighted fluid attenuated inversion recovery sequence showed hyperintense changes in the right mesial temporal lobe, the posterior limb of the right internal capsule, and the right lateral geniculate body (LGB), being consistent with right AChA territory infarction (Fig. 2). The patient was given an antiplatelet agent and discharged with partial improvement of his symptoms. He was then referred to another hospital for right carotid endarterectomy. Four years later, repeat perimetry showed no demonstrable visual field defect in the right eye and marked decrease of visual field defect in the left eye.

## 3. Discussion

The AChA is a small artery that usually originates from the internal carotid artery above the origins of the ophthalmic and posterior communicating arteries.<sup>1</sup> It may also exist as a branch of the middle cerebral or posterior communicating arteries.<sup>4</sup> The AChA generally supplies the lateral portion of the optic tract, the mesial temporal lobe, the cerebral peduncle, the medial portion of the globus pallidus, the lateral thalamus, the posterior two-thirds

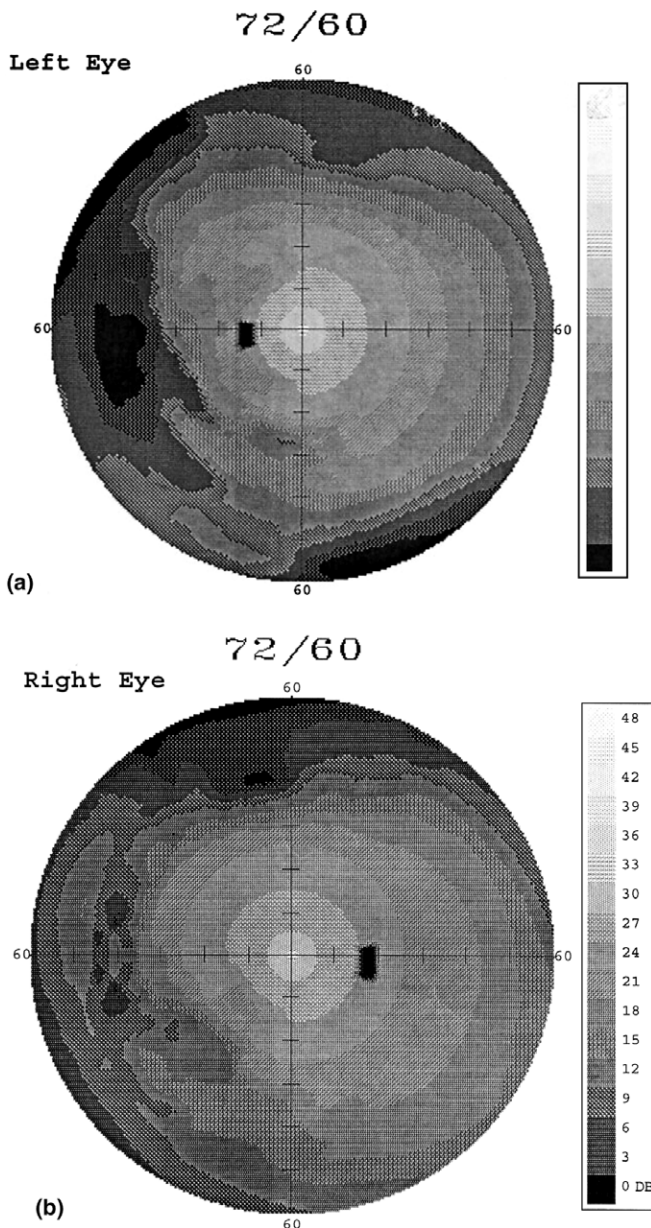


Fig. 1. Automated perimetry showed a left incongruent homonymous hemianopia predominantly affecting lower quadrant with central sparing. Left eye (a); right eye (b).

of the posterior limb of the internal capsule, the lateral half of the LGB, and the choroid plexus.<sup>4–6</sup> The diagnosis of AChA territory infarction is made based on the specific vascular involved, particularly the posterior two-thirds of the posterior limb of the internal capsule, not on occlusion of the AChA by angiography, because a patent AChA does not exclude the diagnosis.<sup>2,5,6</sup> In our patient, the infarcted areas were all within the vascular territory of the AChA.

Homonymous upper quadrantanopsia, hemianopsia, and upper and lower quadrant sectoranopsia have all been observed in patients with AChA territory infarction.<sup>1</sup> The variability of visual dysfunction observed in AChA syndrome is attributable to the variable involvement of the

optic tract, the lateral half of the LGB, and the optic radiation.<sup>1,2</sup> Optic tract infarction should produce an incongruent homonymous hemianopsia, whereas ischaemia of the optic radiation could lead to a congruent homonymous hemianopsia, possibly sparing the macula.<sup>1,3</sup> Involvement of the optic radiation is considered the most common cause of the visual abnormality found in patients with AChA territory infarction.<sup>1</sup> However, this pattern of visual abnormality is often transient because of the rich collateral circulation.<sup>1</sup> Frisen et al. was the first to describe a patient with a homonymous defect in the upper and lower visual fields sparing the horizontal meridian and concluded that the visual pathway was damaged within that part of the LGB supplied by the AChA.<sup>7</sup> This abnormality was named quadruple sectoranopsia.<sup>7</sup> A superior quadrantanopsia with macular sparing, attributed to LGB involvement in AChA territory infarction, has been reported.<sup>1,8</sup> It has been assumed that the representation of each part of the visual field within the LGB could explain the visual field patterns observed. The lower retinal quadrant occupies the anterolateral part of the LGB, the upper retinal quadrant occupies the anteromedial part of the LGB, and the macula is represented in the posterior pole of the LGB.<sup>9</sup> Moreover, the different patterns of visual field defect in LGB infarction may be explained by its dual blood supply from the AChA and from the lateral choroidal artery.<sup>8,9</sup> The visual field defects caused by vascular lesions in the LGB may be incongruent or moderately to completely congruent.<sup>8–10</sup> The visual field defect observed in our patient, the left incongruent homonymous hemianopsia with central sparing, could be the result of ischaemia involving the posterior optic tract, the early optic radiation, or the LGB, in isolation or in various combinations.

Although infarctions in the AChA territory usually result from small-vessel disease, they sometimes occur in association with lesions of the proximal internal carotid artery.<sup>11</sup> Our patient, who had two episodes of unsteady gait along with dysarthria and right periorbital pain, may have had ischaemic episodes as the result of hypoperfusion produced by the severe stenosis of the right internal carotid artery. The ischaemic lesion in the posterior limb of the internal capsule usually causes motor dysfunction, but not always, because of rich collateral circulation from neighbouring arteries.<sup>3</sup>

This patient, who had an AChA territory infarction, was unaware of his visual symptom throughout his illness. Upon review of the literature, the majority of patients with AChA territory infarction who presented with motor dysfunctions were unaware of their visual symptoms.<sup>1,2,7,8</sup> Visual abnormalities were found only by formal visual field examination, and a feature common to all visual abnormalities was the sparing of central vision.<sup>1,7,8</sup> Thus, it can be stated that the visual field defects caused by vascular lesions in the territory of the AChA can easily be overlooked and the incidence underestimated in the presence of intact central vision, particularly if there are no prominent motor and sensory symptoms. This inconspicuous of visual field

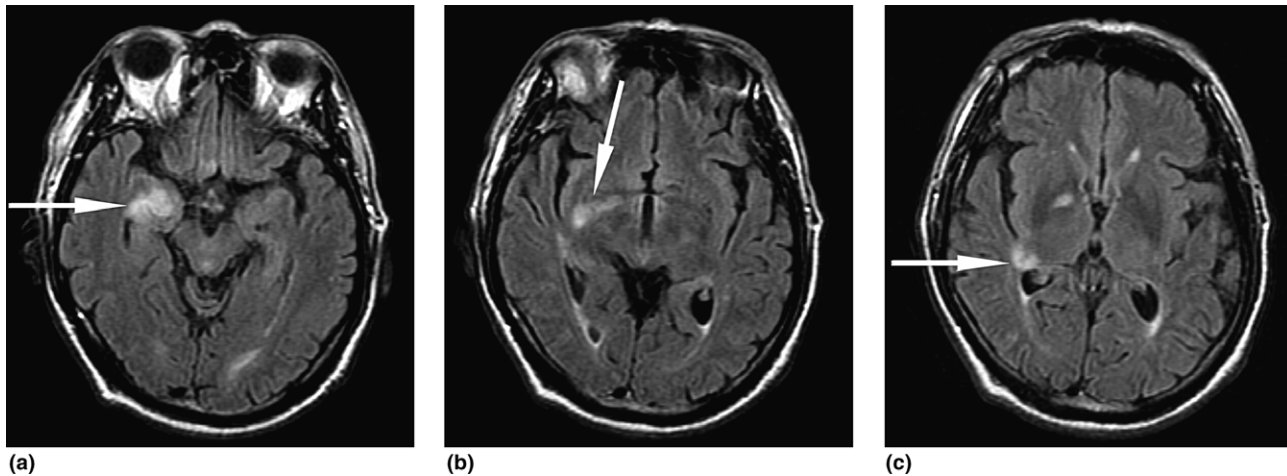


Fig. 2. Axial T2-weighted fluid attenuated inversion recovery images of the brain at the level of the midbrain (a), hypothalamus (b), and thalamus (c). Hyperintense signal change is indicated by white arrows: in (a), the right mesial temporal lobe; in (b), the posterior limb of the right internal capsule; in (c), the right lateral geniculate body.

defect could increase the danger of driving, the risk of falls, and occupational hazards. Therefore, it may be recommended that, in all patients with image findings of AChA territory infarction, a formal visual field assessment is necessary.

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## Carbamyl phosphate synthase deficiency: Diagnosed during pregnancy in a 41-year-old

G. Eather<sup>a</sup>, D. Coman<sup>b,c</sup>, C. Lander<sup>a</sup>, J. McGill<sup>b,c,\*</sup>

<sup>a</sup> Department of Neurology, The Royal Brisbane Hospital

<sup>b</sup> Department of Metabolic Medicine, The Royal Children's Hospital, Herston Road, Herston, Brisbane 4029, Australia

<sup>c</sup> Department of Paediatrics and Child Health, University of Queensland

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\* Corresponding author. Tel.: +61 7 3636 8111; fax: +61 7 3636 1860.  
E-mail address: jim\_mcgill@health.qld.gov.au (J. McGill).