

[ CASE REPORT ]

# Transient Hemichorea-hemiballism Induced by a Combination of Postprandial Hypotension and Severe Stenosis of the Innominate Artery Concomitant with Left Carotid Occlusion

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## Abstract:

Hemichorea-hemiballism (HCHB) due to transient ischemic attacks (TIAs) is rare. An 83-year-old woman had repeated episodes of right-sided HCHB for 3 months. Magnetic resonance (MR) angiography demonstrated occlusion of the left carotid and middle cerebral arteries and severe stenosis of the innominate artery, and 24-h ambulatory blood pressure monitoring showed a blood pressure decrease of >20 mmHg after each meal. We speculated that HCHB developed as TIAs due to hemodynamic failure in the left cerebral hemisphere, caused by a combination of severe stenosis of the innominate artery concomitant with occlusion of the left carotid and middle cerebral arteries as well as postprandial hypotension.

**Key words:** transient ischemic attack, hemichorea-hemiballism, postprandial hypotension, innominate artery stenosis,  $\alpha$ -glucosidase inhibitors

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## Introduction

Hemichorea-hemiballism (HCHB) is an involuntary movement disorder characterized by incidental acute twisting or flinging unilateral movements of a limb caused by stroke or nonketotic hyperglycemia (1). Limb shaking, a widely known involuntary movement that occurs in transient ischemic attack (TIA), is a smaller-scale involuntary movement than HCHB, described as “trembling, shaking, twisting, drawing up or moving irregularly” (2). Limb shaking is considered to be the most common TIA-induced involuntary movement, but HCHB due to TIAs is very rare, except in cases of Moyamoya disease (3).

We herein report an unusual case of paroxysmal HCHB resulting from TIAs induced by postprandial hypotension

(PPH).

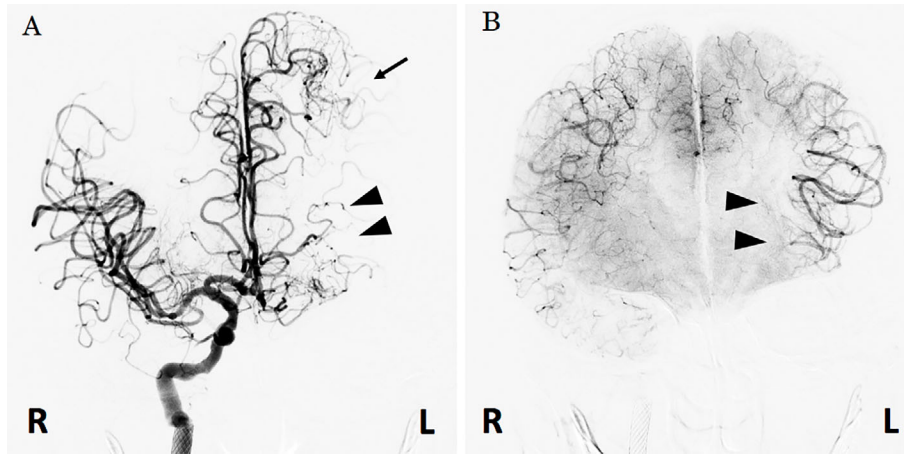
## Case Report

An 83-year-old woman complained of repeated episodes of coarse, involuntary movements in the right arm, lasting for several minutes, for the past 3 months. She was receiving aspirin (200 mg/day) for a history of symptomatic right carotid stenosis revascularized by stent placement 4 years earlier, with occlusion of the contralateral internal carotid and middle cerebral arteries. Cerebral angiography at that time had shown collateral blood flow from the right internal carotid artery (ICA) through the anterior communicating artery to the left anterior cerebral artery (ACA). The left ACA had spread perforating branches to the caudate nucleus and globus pallidus and maintained blood flow to the cortex of

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**Figure 1.** Digital subtraction cerebral angiography, antero-posterior view of right common carotid injection. The arterial phase (A) and the capillary phase (B). A: Collateral flow from the right ICA through the AcomA to the left ACA. The left ACA spread perforating branches to the caudate nucleus and globus pallidus (arrowheads) and maintained blood flow to the cortex of the left hemisphere by leptomeningeal collaterals (arrow). B: The left ACA did not sufficiently feed the left putamen and posterior limb of the internal capsule, resulting in a filling delay (arrowheads). AcomA: anterior communicating artery, ACA: anterior cerebral artery, ICA: internal carotid artery

the left hemisphere via leptomeningeal collaterals (Fig. 1A). Common carotid angiography on the occluded side had confirmed the ICA C2 portion via the ipsilateral ophthalmic artery. Left posterior cerebral artery blood flow through the posterior communicating artery additionally fed the left temporal cortex.

The patient had hypertension controlled with losartan (25 mg/day), a history of 82.5 pack-years of smoking, and the intake of nitrate medications for angina pain. There was no history of dyslipidemia, and her low-density lipoprotein cholesterol level was 91 mg/dL. Fasting and postprandial plasma glucose values were within the normal ranges, and the hemoglobin A1c content according to the NGSP method was 6.0%. She had right renal atrophy due to renal artery stenosis with a low urinary protein value and a stable serum-creatinine level of approximately 1.3 mg/dL. On the premorbid Mini-Mental State Examination (MMSE), she had a score of 19 but had no difficulties with activities of daily living. She had been taking magnesium oxide (1,000 mg/day) for constipation, which occurred approximately once every 2 days. Her electroencephalogram findings were normal, and antiepileptic agents prescribed by another doctor had proven ineffective.

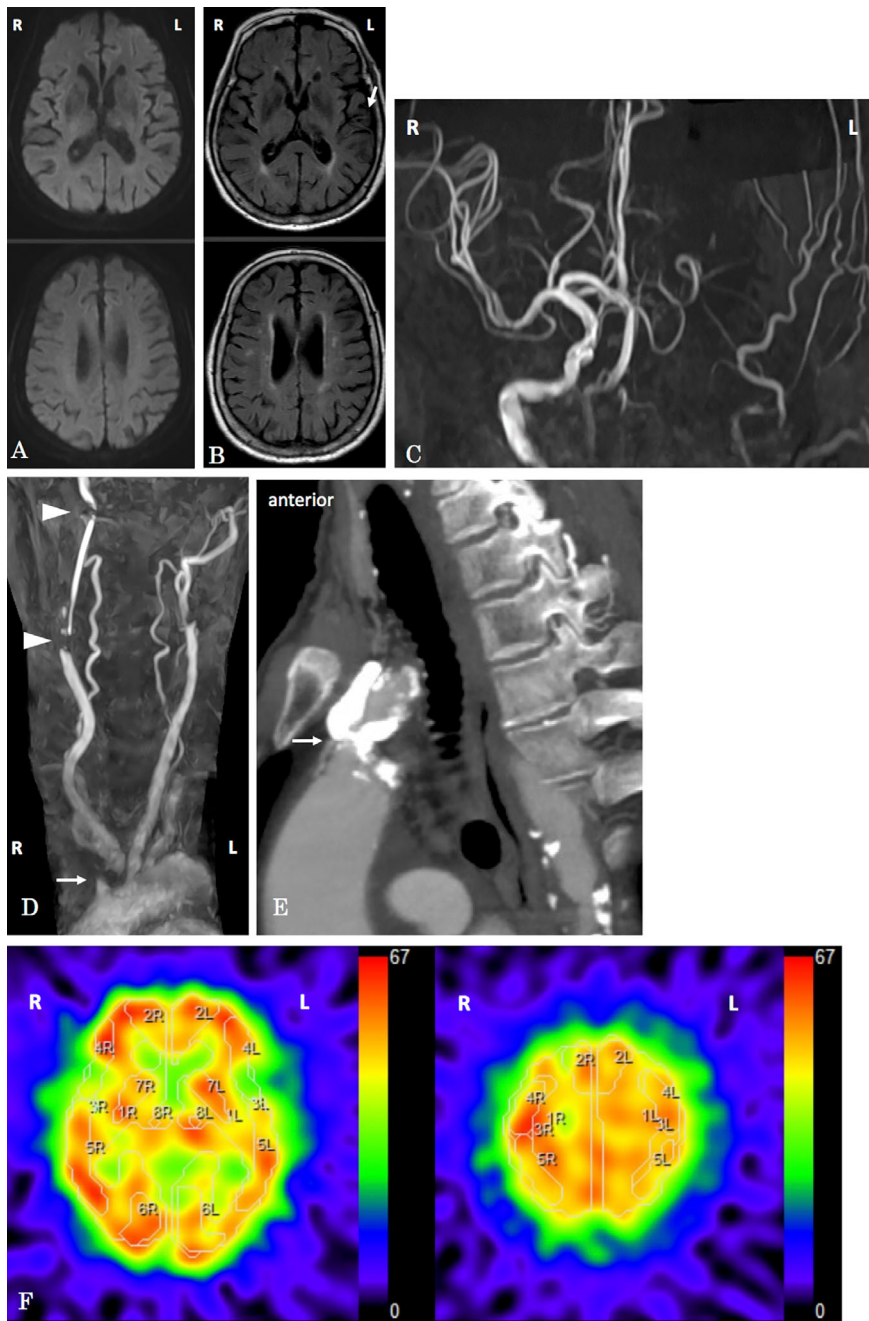
After referral to our hospital, she suddenly developed transient right hemiparesis and aphasia in the waiting room approximately one hour after snacking. She was admitted to our hospital for presumed TIAs. On admission, she had normal deep tendon reflexes and did not exhibit any neurological deficits, including cerebellar ataxia. However, she did show a marked difference in the blood pressure (BP) in the bilateral arms (129/109 mmHg on the right side and 188/49 mmHg on the left). In addition, her BP repeatedly fell by approximately 30 mmHg with postural changes from the supine to the standing position. The hypotension associated

with the position change was not persistent, and no manifestation of involuntary movement occurred. There were no other abnormal physical findings.

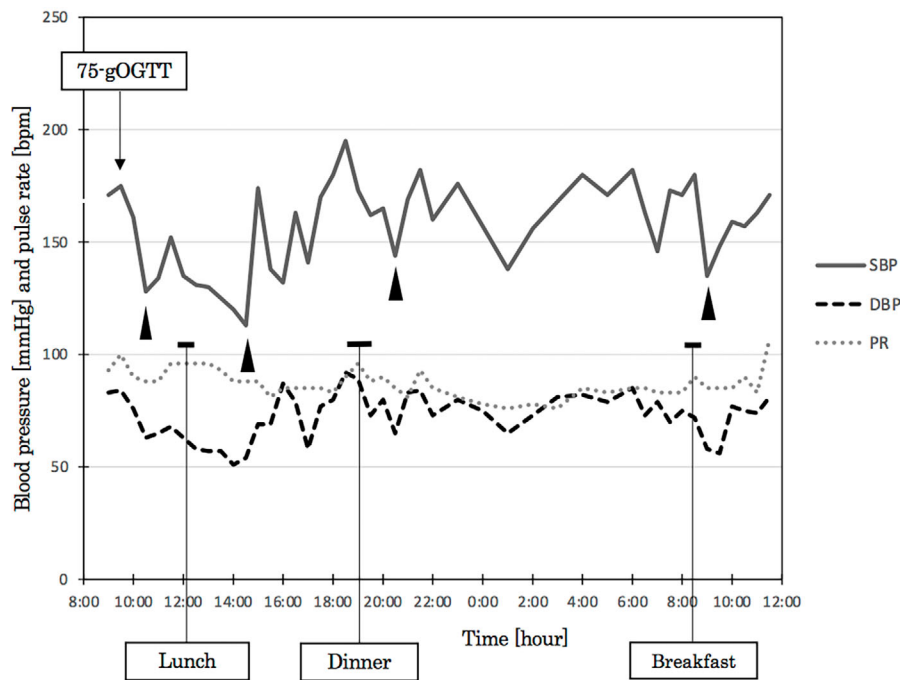
During her hospital stay, she occasionally developed paroxysmal dizziness with twisting or writhing of the right hand and flinging movements of the right arm and leg, consistent with HCHB, one to two hours after eating (see Video Segment 1). Diffusion-weighted magnetic resonance imaging showed no high-signal lesions (Fig. 2A). Fluid attenuated inversion recovery (FLAIR) imaging showed no obvious old infarctions, but hyperintense vessel signs were present around the left temporal and insular lobes (Fig. 2B). On head magnetic resonance angiography (MRA), the left internal carotid and middle cerebral arteries were not visualized (Fig. 2C). Thoracic MRA and contrast-enhanced computed tomography revealed heavily calcified, severe ostial stenosis of the innominate artery (Fig. 2D, E). Brain single-photon emission computed tomography 123I-IMP in the resting state showed no apparent cortical hypoperfusion (Fig. 2F).

Twenty-four-hour ambulatory BP monitoring showed a BP decrease of >20 mmHg after each meal (Fig. 3). In addition, her coefficient of variation of R-R intervals decreased to 2%. We speculated that HCHB had developed from TIAs due to hemodynamic failure in the left cerebral hemisphere caused by severe stenosis of the innominate artery, which was the main source of blood flow to the left cerebral hemisphere, and PPH.

The stenotic lesion of the innominate artery was hard to revascularize endovascularly due to its heavy calcification, and surgical revascularization, such as bypass surgery, was too invasive to be performed in this patient due to her multiple comorbidities. We therefore prescribed diet therapy to reduce the PPH, including dividing meals into smaller portions, enacting carbohydrate restriction, and encouraging



**Figure 2.** Axial diffusion-weighted brain MRI (A), axial FLAIR image (B), head MRA (C), MRA from the aortic arch to the neck (D), CTA of the aortic arch (E), and brain SPECT 123I-IMP in the resting state (F). A: Diffusion-weighted MRI showed no high-signal lesions. B: FLAIR images did not reveal any obvious old infarctions but showed hyperintense vessel signs around the temporal and insular lobes (arrow). There were diffuse mild atrophic changes and several white matter hyperintensities of the bilateral cerebral hemispheres. C: On head MRA, the left internal carotid and middle cerebral arteries are not visualized. D: MRA demonstrating an occluded left internal carotid artery and severe stenosis of the innominate artery (arrow). The right carotid artery had previously undergone revascularization (between two arrowheads). Carotid duplex ultrasonography showed no significant in-stent restenosis (images not shown). E: CTA demonstrated heavily calcified, severe ostial stenosis of the innominate artery. F: Brain SPECT showed no apparent cortical hypoperfusion in the MCA, ACA, or basal ganglia regions (an acetazolamide challenge was not performed). The cerebral blood flow of the MCA, ACA, and basal ganglia regions was 49.11 and 51.36 mL/100 g/min, 49.87 and 48.39 mL/100 g/min, and 52.26 and 50.08 mL/100 g/min on the left and right sides, respectively. ACA: anterior cerebral artery, CTA: computed tomography angiograph, FLAIR: fluid attenuated inversion recovery, MCA: middle cerebral artery, MRA: magnetic resonance angiography, MRI: magnetic resonance imaging, SPECT: single-photon emission computed tomography



**Figure 3.** A 24-h ambulatory blood pressure monitoring chart showing that blood pressure decreased by 20-40 mmHg after a 75-g OGTT and each meal (arrowheads: nadirs of the blood pressure drops). DBP: diastolic blood pressure, PR: pulse rate, SBP: systolic blood pressure, 75-g OGTT: 75-g oral glucose tolerance test

green tea consumption after each meal as a route of intake caffeine. In addition, we instructed her to lie down for approximately one hour after each meal.

The PPH finally resolved a few days after the introduction of an alpha-glucosidase inhibitor (GI) and 0.3 mg of voglibose before each meal. The average BP 2 h after lunch following medication intake decreased by approximately 20 mmHg; this was attributed to reduced BP fluctuation compared to the pretreatment average BP, which had decreased by approximately 40 mmHg after lunch previously. Treatment with low-dose oral amlodipine was initiated to control the increased BP. Thereafter, she had no TIAs or paroxysmal HCHB for over 2.5 years of follow-up.

## Discussion

This case was characterized by an unusual manifestation of TIAs when severe stenosis of the innominate artery and occlusion of the left carotid and middle cerebral arteries, combined with PPH, resulted in insufficient cerebral blood flow to the left cerebral hemisphere, causing HCHB. Administration of an alpha-GI improved the PPH without any recurrence of TIAs.

Limb shaking is considered to be the most common TIA-induced involuntary movement, but HCHB due to TIAs is very rare, except for in cases of Moyamoya disease. Only six other studies of hemiballism or HCHB caused by TIA with cerebral large-vessel steno-occlusive disease have been reported (Table 1) (4-8). In the seven total cases, involuntary movements occurred on the contralateral side of a large-

vessel steno-occlusive lesion. The present case was, however, the only case in which the large-vessel steno-occlusive lesion ipsilateral to the affected half of the body was simultaneously associated with the development of involuntary movements. While HCHB is thought to be derived from disruptions of the indirect pathway by lesions of the subthalamic nucleus, lesions in other parts of the loop (such as the putamen, motor cortex, and others) may also be causative (1). In the present case, the severely stenosed innominate artery was the main source of blood flow to the left cerebral hemisphere due to the total occlusion of the left carotid and middle cerebral arteries and directly caused TIA with HCHB combined with PPH. The previous cerebral angiography procedure showed that the perforating branches and leptomeningeal collaterals from the left ACA did not sufficiently feed the left putamen and posterior limb of the internal capsule, resulting in a filling delay (Fig. 1B). This probably resulted in insufficient collateral blood flow with subsequent impairment of the indirect pathway connecting to the cortico-basal ganglia loop.

TIA caused by PPH has been reported with autonomic dysfunction, such as neurodegenerative diseases, diabetes mellitus, and large-vessel steno-occlusion (Table 2) (9-15). In six of the nine total reported cases (including the present one), orthostatic hypotension partly associated with neurodegenerative diseases or autonomic neuropathy (idiopathic or diabetic) were comorbid conditions associated with PPH. In the other three cases, PPH occurred in postgastrectomy patients, likely due to insufficient compensation for the decrease in circulating blood flow due to increased gastrointes-

**Table 1. HBHC Cases Caused by TIA with Cerebral Large-vessel Steno-occlusive Disease.**

Reference	Age / Sex	Lesion site	Location of involuntary movements	Involuntary movements
(4)	72 / M	Bil ICA stenosis	Bil arms and legs	HBHC
(5)	62 / M	Rt MCA stenosis	Lt arm and leg	Hemiballism
	61 / M	Rt MCA stenosis	Lt arm and leg	Hemiballism
(6)	71 / M	Lt MCA stenosis	Rt arm and leg	Hemiballism
(7)	51 / F	Rt MCA stenosis	Lt arm and leg	HBHC
(8)	44 / F	Rt MCA occlusion, Rt ACA stenosis	Lt arm and leg	Hemiballism
Present case	83 / F	Innominate artery stenosis (Lt ICA · MCA occlusion)	Rt arm and leg	HBHC

ACA: anterior cerebral artery, Bil: bilateral, HBHC: hemiballism-hemichorea, ICA: internal carotid artery, Lt: left, MCA: middle cerebral artery, Rt: right, TIA: transient ischemic attack

**Table 2. Cases of Transient Ischemic Attack Caused by PPH.**

Reference	Age / Sex	Symptoms	Cerebrovascular steno-occlusive lesion	causative diseases	Treatment
(9)	72 / M	Blindness and severe periorbital pain in the Rt eye	Rt intracranial ICA stenosis	History of pyloroplasty and vagotomy	Anticoagulant therapy, Dividing meals into smaller portions
(10)	78 / M	Rt hemiparesis Dysarthria Aphasia	Lt ICA occlusion	OH	Caffeine, Lying down after each meal
(11)	78 / M	Rt hemiparesis, Aphasia	Lt ICA occlusion	History of gastrectomy	Lying down after each meal
(12)	75 / F	Rt hemiparesis Disorientation Dysarthria	No lesion	OH Parkinsonism	Caffeine, Lying down after each meal
	61 / M	Rt hemiparesis Syncope Dysarthria	No lesion	OH Multiple system atrophy	No specific treatment
(13)	76 / M	Rt hemiparesis	Lt ICA occlusion	Diabetes	Alpha-GI (voglibose)
(14)	61 / F	Rt-sided limb shaking Lightheadedness	Lt ICA occlusion	Idiopathic autonomic neuropathy	Dividing meals into smaller portions, Restricting carbohydrates Avoiding prolonged standing for an hour after each meal
(15)	82 / F	Rt hemiparesis Consciousness disturbance	Lt ICA stenosis	History of gastrectomy	Alpha-GI (voglibose)
Present Case	83 / F	Rt-sided HBHC Rt hemiparesis Aphasia	Innominate artery stenosis (Lt ICA · MCA occlusion)	OH PPH	Dividing meals into smaller portions Restricting carbohydrates Caffeine, Lying down after each meal Alpha-GI (voglibose)

CAS: carotid artery stenting, CEA: carotid endarterectomy, HBHC: hemiballism-hemichorea, ICA: internal carotid artery, Lt: left, MCA: middle cerebral artery, OH: Orthostatic hypotension, PPH: postprandial hypotension, Rt: right

tinal blood flow during digestion. In the present case, we detected orthostatic hypotension, constipation, and an impaired coefficient of R-R interval variation as autonomic neuropathy associated with aging. However, her HbA1c (NGSP) value remained below 6.0%, her fasting and postprandial blood glucose levels were normal at the time of the disease onset, and there were no abnormalities in glucose tolerance that would lead to a diagnosis of diabetes mellitus. We concluded that her ability to regulate BP was impaired due to the autonomic dysfunction associated with

aging and systemic atherosclerosis, including the aortic arch and its major branches, coronary arteries, and renal and other arteries. Because PPH was combined with latent orthostatic hypotension, the patient developed severe hypotension sufficient to induce cerebral ischemia when sitting or standing after eating.

Older adults may sometimes lack a sufficient increase in sympathetic activity responding to an oral glucose load to compensate for the normal physiologic postmeal decrease in BP (16). Alpha-GIs delay digestion and glucose absorption,

preventing PPH by ablating the subsequent increase in gastrointestinal blood flow (13, 15-17).

The present findings support the notion that patients with recurrent postprandial HCHB should be evaluated for autonomic dysfunction and cerebral large-vessel steno-occlusive diseases.

**The authors state that they have no Conflict of Interest (COI).**

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### Ethical approval statement/IRB approval number

This study was approved by the Ethics Committee of the University of Tsukuba Hospital (approval no. R01-216). Informed consent was obtained from the patient.

### Declaration of conflicting interests

The authors declare no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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