of the hyperautonomic episodes. Replacement with 100 mg labetalol twice daily led to reduction in the frequency of events to about one a day. Subsequent increase of the medication to 200 mg twice daily resulted in a marked decrease to less than one paroxysmal sympathetic storm over several days. At the time of discharge, the patient had returned to his preadmission baseline.

The current observations lend support to the prevailing view that paroxysmal sympathetic storm may represent disruption of autonomic function in the diencephalon and brainstem. Bullard has proposed that the clinical syndrome may be the result of a release phenomenon within the brainstem and/or diencephalon from loss of overriding cortical or subcortical inhibition.[3] More recent case studies suggest localisation to the central sympathoexcitatory regions including the paraventricular hypothalamic nucleus, lateral periaqueductal grey matter, lateral parabrachial nucleus, or rostral ventrolateral medulla.[3] Compromised autonomic neuronal integrity centrally is not surprising in the setting of infection after traumatic brain injury.

Various medications can potentially be used in managing central sympathetic storm. Imidazoline agonists and specific α2 adrenoceptor antagonists, such as clonidine and methyldopa,[5] have recently been shown to have sympathoinhibitory actions centrally within the rostral ventrolateral medulla. These agents have so far been used in the treatment of essential hypertension, tetanus, or autonomic dysreflexia. β Blockers such as propranolol however, have long been the mainstay of treatment of the hypertension, tachycardia, and hyperpyrexia associated with paroxysmal sympathetic storm.[6 7] This non-selective β adrenergic antagonist acts through inhibition of peripheral catecholamine activity, and being highly lipophilic, may also exert central effects through membrane stabilisation or receptor blockade. Moreover, propranolol may reduce sustained muscle contraction.[7] Taken together, these findings suggest that non-selective β receptor antagonism is sufficient to inhibit the clinical manifestations of diencephalic seizures.

The present case suggests that β1 receptor antagonism alone is not sufficient to treat hyperautonomia during paroxysmal sympathetic storm. This patient was initially placed on starting doses of metoprolol, a selective β1 antagonist, with little clinical effect in controlling the frequency of the autonomic attacks; however, labetalol, an α1 and β1-β2 adrenergic receptor antagonist did lead to an observable decline in symptoms. Both sympatholytic agents were given at doses typically used in initiating treatment of systemic hypertension, suggesting that the observed response seen with labetalol could not be explained solely by a dosage phenomenon. Prior studies also demonstrate that small amounts of propranolol (20 mg four times a day) can achieve similar responses to those seen with labetalol,[6 7] further arguing against a dose dependent effect. Thus, at a minimum, either α1 or β2 receptor blockade, likely in addition to β1 blockade, is necessary in the treatment of paroxysmal sympathetic storm.

The discrepancy in response between metoprolol and labetalol could result from their different effects on the cardiovascular system or CNS. The β1-β2 adrenergic receptor blockade by labetalol decreases blood pressure and heart rate through negative inotropic and chronotropic effects, and by inhibiting renin release. In addition, labetalol has vasodilator properties resulting from α1 blockade and partial β2 agonism. These reduce peripheral vascular resistance, blood pressure, and coronary vascular resistance, a potential advantage over other β blockers. Alternatively, differences in central activity may explain the increased efficacy of labetalol over metoprolol. As both agents are lipophilic, their central access should not differ significantly; rather, differences in receptor antagonism (β1 versus α1, β1, β2) would more likely explain the therapeutic discrepancy. As proposed with propranolol,[4] inhibition of β2 receptors by labetalol may exert a stabilising effect within the CNS through indirect inhibition of sympathetic nerve activity.

In the present case, we report the use of labetalol as an alternative agent in the treatment of paroxysmal sympathetic storm. It likely exerts both a central and peripheral blockade of α1 and β adrenergic receptors to produce inhibition of autonomic dysregulation. The clinical ineffectiveness of metoprolol further suggests a necessary role for β2 and/or α1 receptors in the clinical presentation of paroxysmal sympathetic storm. Labetalol may prove an alternative equal to or better than morphine in the treatment of these spells, especially when addiction and dependency are of concern.

D DO
V L SHEEN
E BROMFIELD
Departments of Neurology, Brigham and Women's Hospital, 75 Francis Street, Boston, MA 02115, USA and Massachusetts General Hospital, Harvard Medical School, Boston, MA, USA

Correspondence to: Dr E Bromfield
ebromfield@partners.org

1 Penfield W, Jasper H. *Epilepsy and the functional anatomy of the human brain.* 1st ed. Boston: Little Brown, 1954.
2 Boeve BF, Wijdicks EF, Benarroch EE, *et al*. Paroxysmal sympathetic storms (diencephalic seizures) after severe diffuse axonal head injury. *Mayo Clin Proc* 1998;**73**:148–52.
3 Bullard DE. Diencephalic seizures: responsiveness to bromocriptine and morphine. *Ann Neurol* 1987;**21**:609–11.
4 Head GA. Central imidazoline- and α2-receptors involved in the cardiovascular actions of centrally acting antihypertensive agents. *Ann NY Acad Sci* 1999;**881**:279–86.
5 Jenkins WL, Clark DR. A review of drugs affecting the heart. *J Am Vet Med* Assoc 1977;**171**:85–92.
6 Meythaler JM, Stinson AM. Fever of central origin in traumatic brain injury controlled with propranolol. *Arch Phys Med Rehabil* 1994;**75**:816–18.
7 Sneed RC. Hyperpyrexia associated with sustained muscle contractions: an alternative diagnosis to central fever. *Arch Phys Med Rehabil* 1995;**76**:101–3.

Moyamoya disease presenting with singing induced chorea

Moyamoya disease is a relatively uncommon, chronic cerebral vasculopathy of unknown aetiology that is characterised by unilateral or bilateral stenosis or occlusion of the proximal portion of the carotid arteries, together with an abnormal vascular network at the base of the brain. Most childhood cases manifest with the signs and symptoms of cerebral ischaemia or infarction, whereas intracerebral haemorrhage prevails in adults.[1 2] We describe here a case of moyamoya disease in a 29 year old multiparous woman, who presented with involuntary limb movements induced by singing.

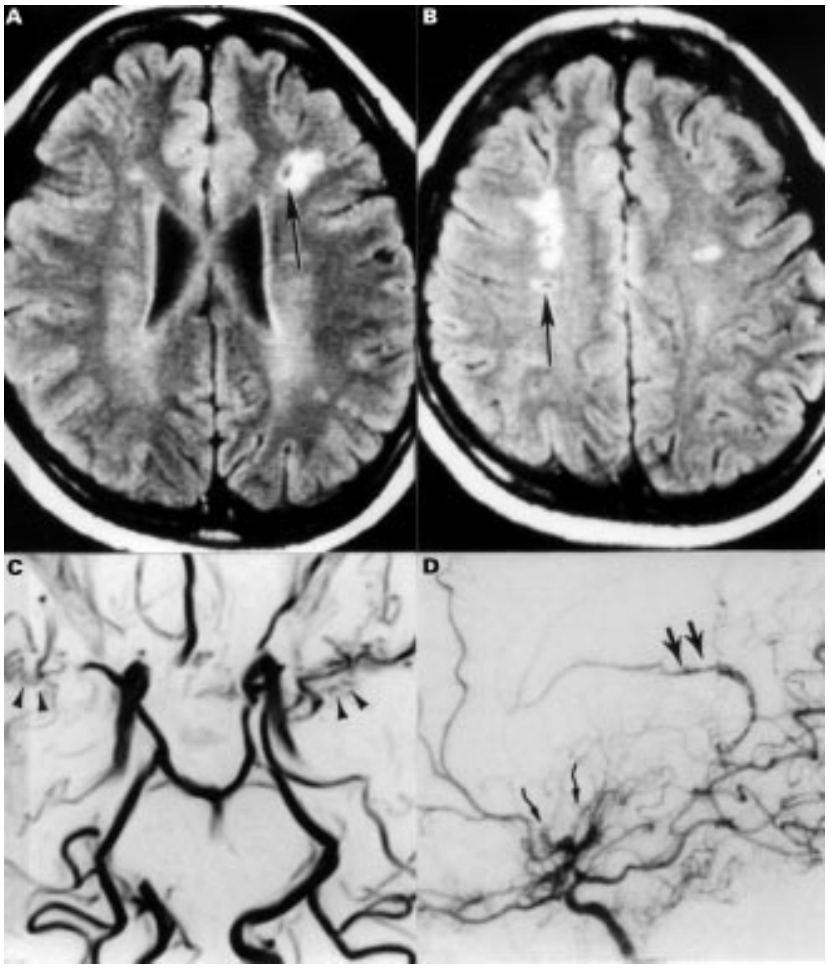
A 29 year old woman, gravida two, para two, presented to the neurological outpatient clinic at Chungbuk National University Hospital with recurrent episodes of brief involuntary movements affecting her left hand and arm. The movements were characterised as unilateral, brief, coarse, irregular, and wavering. There was no history of neuroleptic drug therapy, or family history of involuntary movement.

General physical, neurological, and neuropsychological examinations were unremarkable. Baseline blood tests, ECG, and chest radiography all yielded normal results. The episodes of the patient's involuntary movements were unique, in that they usually appeared while she was opera type singing in a choir at church. They were also occasionally provoked by some conditions of hyperventilation such as blowing to cool hot soup, or blowing the dust off a table. This suggested an underlying ischaemic pathophysiology and prompted us to investigate changes in brain vasculature and parenchyma. The short lived choreiform movements were usually preceded by a tingling sensation in her left hand, which occasionally extended to the left leg.

An EEG between ischaemic episodes disclosed diffuse slow waves bilaterally over the hemispheres; these slow waves increased as "build up" with the appearance of delta waves during hyperventilation. Magnetic resonance imaging showed areas of high signal intensity in both frontal subcortical regions, suggestive of focal ischaemic lesions (fig A and B). We determined the patient's cerebral vascular reserve using technetium-99m-HMPAO brain SPECT with acetazolamide challenge. This demonstrated a decreased vascular reserve in both frontal and temporal lobes, as well as in the basal ganglia. Magnetic resonance angiography and subsequent four vessel angiography showed nearly complete obstruction of the terminal portion of each internal carotid artery and the outline of a moyamoya network (fig C and D). Staged encephaloduroarteriosynangiosis was performed on the left and right sides, 1 week apart, resulting in an eventual amelioration of the patient's involuntary movements.

Chorea is one of the rarer, although acknowledged, presenting features of moyamoya disease; chorea is usually observed in children.[3 4] It is suggested that about 6% of patients with moyamoya disease have chorea.[5] Other types of involuntary movements have been described in patients with moyamoya disease: Valsalva related seizures,[6] recurrent episodes of carpopedal spasm,[7] recurrent torticollis,[8] and limb shaking transient ischaemic attack.[9] Hemichorea is characterised by unilateral, brief, coarse, irregular, wavering, involuntary movements, and is usually caused by some asymmetric, focal brain lesion. The clinical presentation of our patient was associated with opera type singing.

Singing requires both hyperventilation and the breath holding Valsalva's manoeuvre. Hyperventilation causes an increase in arterial oxygen tension, which subsequently causes vasoconstriction, which, in turn, reduces blood flow. In addition, Valsalva's manoeuvre increases cerebral venous pressure, which then increases intracranial blood volume and intracranial pressure, thereby reducing the arterial perfusion pressure. Thus, in those regions of the basal ganglia and cortex that are already critically perfused, hyperventilation and Valsalva's manoeuvre can easily lead to transient ischaemic insult, which may be clinically manifested by involuntary movements. It seems likely that hyperventilation and breath holding act synergistically to reduce brain perfusion. In this patient, the hemichoreic episodes were attributed to hypoperfusion of the contralateral cerebral hemisphere, and not to epilep-



(A and B) FLAIR axial images, showing bilateral focal ischaemic lesions in the frontal white matter. Low signal intensities surrounded by hyperintense rims are chronic lacunae (arrows) (C) Time of flight MR angiography, indicating that both middle cerebral arteries were unidentifiable. Note multiple tortuous flow signals, suggestive of moyamoya vessels (arrowheads). (D) Right internal carotid angiogram, demonstrating middle cerebral artery occlusion, moyamoya vessels (curved arrows), and the leptomeningeal collateral blood flow from the posterior circulation (straight arrows).

togenic activity. Staged left and right encephaloduroarteriosynangiosis, using a frontal branch of the superficial temporal artery, was carried out, 1 week apart. This procedure eventually ameliorated the patient's choreic movements.

Chorea is not unusual in moyamoya disease. However, the causes of chorea are manifold and careful neuroradiological and clinical evaluation is required to distinguish them.[10] Our findings emphasise that moyamoya disease should be included in the differential diagnosis of adult onset chorea. Recognition of this uncommon form of occlusive carotid disease is important in the early diagnosis and proper management of neurological deficits.

This study was partly supported by a Clinical Research Fund from Chungbuk National University Hospital.

S-H HAN
Department of Neurology, Chungbuk National University Hospital, Chungbuk 361–711, Korea

Y-G KIM
Department of Neurosurgery

S-H CHA
Department of Radiology

S-Y CHUNG
Department of Pediatrics, Our Lady of Mercy Hospital, Catholic University Medical College, Inchon, Korea

Correspondence to: Professor Seol-Heui Han
shhan@med.chungbuk.ac.kr

1 Suzuki J, Kodama N. Moyamoya disease: a review. *Stroke* 1983;**14**:104–9.
2 Bruno A, Adams HP Jr, Biller J, *et al*. Cerebral infarction due to moyamoya disease in young adults. *Stroke* 1988;**19**:826–33.
3 Watanabe K, Negoro T, Maehara M, *et al*. Moyamoya disease presenting with chorea. *Pediatr Neurol* 1990;**6**:40–2.
4 Levin SD, Hoare RD, Robinson RO. Childhood moyamoya presenting as dementia: report of a case. *Dev Med Child Neurol* 1983;**25**:794–7.
5 Pavlakis SG, Schneider S, Black K, *et al*. Steroid-responsive chorea in moyamoya disease. *Move Disord* 1991;**4**:347–9.
6 Robertson NP, Compston DAS, Kirkpatrick P. Moyamoya disease presenting as Valsalva related partial seizures. *J Neurol Neurosurg Psychiatry* 1999;**66**:111.
7 Chaudhuri KR, Edwards R, Scott J, *et al*. Adult moyamoya disease. *BMJ* 1993;**307**:852–4.
8 Yasutomo K, Hashimoto T, Miyazaki M, *et al*. Recurrent torticollis as a presentation of moyamoya disease. *J Child Neurol* 1993;**8**:187–8.
9 Ohkawa S, Tabuchi M, Yamadori A. A case of moyamoya-like disease with repetitive TIA on drinking. *Rinsho Shinkeigaku* 1992;**32**:758–62.
10 Pelletier J, Cabanot C, Levrier O, *et al*. Angiodysplasia of moyamoya type disclosed by choreic involuntary abnormal movements during oral contraception. Apropos of 2 cases. *Rev Neurol (Paris)* 1997;**153**:393–7.

Late recurrence of glossopharyngeal neuralgia after IXth and partial Xth nerve rhizotomy: treatment by microvascular decompression

Glossopharyngeal neuralgia, or vagoglossopharyngeal neuralgia as some would prefer,[1] is a rare condition, occurring with a frequency of about 1% of that of trigeminal neuralgia. Medical treatment, particularly with carbamazepine, is usually effective. A significant number of patients do, however, become refractory and go on to surgical treatment. The best established surgical treatment is rhizotomy of the glossopharyngeal and upper vagal nerve roots, which seems to be invariably effective if the diagnosis is correct although it is not without morbidity and even mortality.[2] Late recurrence after such treatment, as described below, has not previously been reported and raises interesting issues of mechanism and method of treatment which are considered in this brief report.

The patient initially presented in 1988 as a 23 year old woman with typical glossopharyngeal neuralgia, experiencing severe intermittent pain in the left side of the throat, the back of the tongue, and the ear. The pain was aggravated by talking and swallowing and relieved, to some degree, by pressure on the left side of the neck. At first there was a good response to carbamazepine. When medication was stopped after several months the pain returned and was less well controlled with a further course of the drug. Neurological examination, CT, and MRI were normal. In 1989 she underwent posterior fossa craniectomy and exploration of the IXth and Xth cranial nerve roots. No lesion, in particular no vascular compression, was identified. The left IXth nerve root and the two uppermost Xth nerve rootlets were divided adjacent to the brain stem. Her postoperative course was uncomplicated and she remained entirely symptom free for over 9 years.

In 1998, now aged 33 years, she developed recurrence of her original pain which she described as essentially identical to that at the initial presentation. Again the pain responded to carbamazepine but required a high dose (1200 mg daily) which was accompanied by troubling side effects (drowsiness and dizziness). In addition she was not completely pain free. Neurological examination and further MRI were normal. In October 1998 a further posterior fossa exploration was carried out. The previously divided nerve roots were identified and the completeness of the initial section confirmed. There was now, however, a large, ectatic vertebral artery to which the proximal ends of the previously sectioned roots were adherent and which was distorting the remaining Xth nerve rootlets and the XIth nerve. A microvascular decompression was carried out with a Teflon patch being placed between the ectatic artery and the normal and previously sectioned nerve roots. The procedure was without complication and the patient has remained well and entirely pain free since that time (18 months).

The first description of glossopharyngeal neuralgia is credited to Weisenberg in 1910, in a patient in whom the pain was secondary to a cerebellopontine angle tumour. The pain is characteristic although two variants have been described; an otitic form with pain predominantly deep in the ear, in the external acoustic meatus, and the mastoid region and an oropharyngeal form in which the pain is experienced in the pharynx, the tonsillar area,