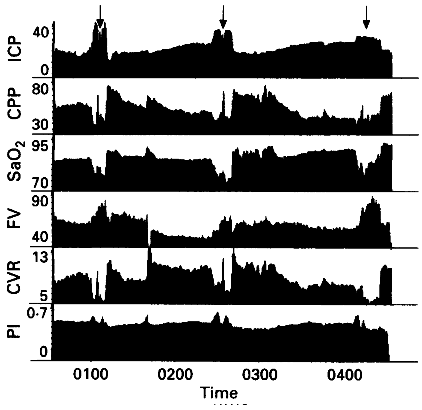

Figure 1 Recording of a four hour period of continuous signal monitoring while the patient was asleep. Three episodes of transient hypoxia are captured (arrowed) during which peripheral saturation fell to 70–75%, ICP rose to 35–40 mm Hg, and CPP fell to 40–45 mm Hg. The increase in FV and fall in calculated CVR are compatible with cerebral vasodilation. ICP = intracranial pressure (mm Hg); CPP = cerebral perfusion pressure (mm Hg); FV = middle cerebral artery flow velocity (cm/s); CVR = cerebral vascular resistance ((mm Hg/s)/cm); PI = pulsatility index of FV.

further arterial desaturation to a mean of 73% occurred with an increased end tidal Pco_2 of 7·8 kPa. These episodes were associated with hypoventilation (reduced abdominal and chest wall movements) without apnoea or airway obstruction. She was given a continuous positive airway pressure device, which provided minor symptomatic relief and improved the mean baseline arterial saturation to 97·5% but failed to abolish the high nocturnal waves of intracranial pressure. A ventriculoperitoneal shunt was therefore inserted and a further period of monitoring undertaken (fig 2). The changes in middle cerebral artery flow velocity and cerebrovascular resistance still occurred with the episodes of desaturation, but the associated increases in intracranial pressure were abolished. At six month follow up her headaches had disappeared, the papilloedema had resolved, and the acuity in the right eye had improved to 6/12.

Several possible aetiological factors for benign intracranial hypertension may have contributed to the visual deterioration in this patient including subclinical cerebral venous thrombosis.[1 2] Although the primary aetiology in our case is not known, raised intracranial pressure occurred during sleep and accompanied episodes of hypoxia and hypercapnia. These were associated with haemodynamic changes compatible with

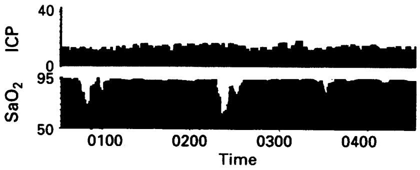

Figure 2 Nocturnal record of ICP and Sao2 after the insertion of a ventriculoperitoneal shunt. The hypoxic events still occur, but the associated increases in ICP have been abolished. ICP = intracranial pressure (mm Hg); Sao2 = peripheral oxygen saturation (%).

cerebral vasodilation. We therefore suggest that nocturnal hypoventilation producing cerebral hypoxia, hypercapnia, and a subsequent rise in intracranial pressure secondary to increased cerebral blood volume contributed to the symptoms. Chronic respiratory disease with severe hypercapnia has long been recognised as a cause of raised intracranial pressure and papilloedema[3] but there were no such features in this case, and although a significantly raised CSF pressure is required for the diagnosis of benign intracranial hypertension, the clinical and radiological features in our patient were typical of this condition.[1] Further, low baseline CSF pressures are often found in patients with chronic benign intracranial hypertension despite persisting papilloedema.[4] Two important points are raised. Firstly, abnormal CSF dynamics require continued observation over several hours as baseline CSF pressure may be normal and waves of raised intracranial pressure transient. Inadequate attention to CSF dynamics may partly explain why isolated CSF pressure estimations do not predict the development of papilloedema and visual deterioration.[5] Secondly, although nocturnal hypoventilation has not been quoted as a contributing factor in benign intracranial hypertension, a relation with raised intracranial pressure has been found. Overnight monitoring of peripheral oxygen saturation may be a useful addition to the investigation of obese patients with symptoms of raised intracranial pressure.

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Hemichorea reversible after operation in a boy with cavernous angioma in the head of the caudate nucleus

Hemichorea and hemiballism point to a structural lesion in the contralateral basal ganglia with a large list of possible causes, including various vascular malformations. Cavernous angiomas are congenital vascular malformations that are occult on conventional angiography (hence "cryptic" vascular malformations (CVMs) but have a characteristic, nearly diagnostic MR image.[1] The definitive diagnosis and distinction from other cryptic vascular malformations depends on histological examination. The clinical manifestations of cavernous angiomas include epilepsy, acute signs secondary to (recurrent) bleeding, and rarely progressive neurological deficit due to expansion of a mass of thrombi within the angioma. With the availability of MRI the number of clinical reports on the subject of CVMs has increased. Recently a case was reported of cavernous angioma in the lentiform nucleus that was the first to present with a movement disorder, in this case focal dystonia. Complete resection was followed by resolution of the symptoms.[2]

We report an 11 year old boy with cavernous angioma in the caudate nucleus, presenting with contralateral hemichorea, evidence of recurrent bleeding, and the disappearance of the hemichorea after surgery.

The boy complained of involuntary movements of the right half of his body including his face, arm and leg, that had suddenly started the week before admission. He could not suppress these movements. There was no family history of neurological disease.

The neurological examination on admission showed continuous, random, jerking movements of the face and extremities on the right side of the body. Muscle strength, sensation, and reflexes were normal.

Brain MRI (figure A) showed a lesion in the head of the caudate nucleus, with the typical aspect of a cavernous angioma.

Two weeks later the boy experienced a sudden deterioration, with involuntary movements of a larger amplitude, more appropriately termed hemiballistic. Surgery was considered appropriate.

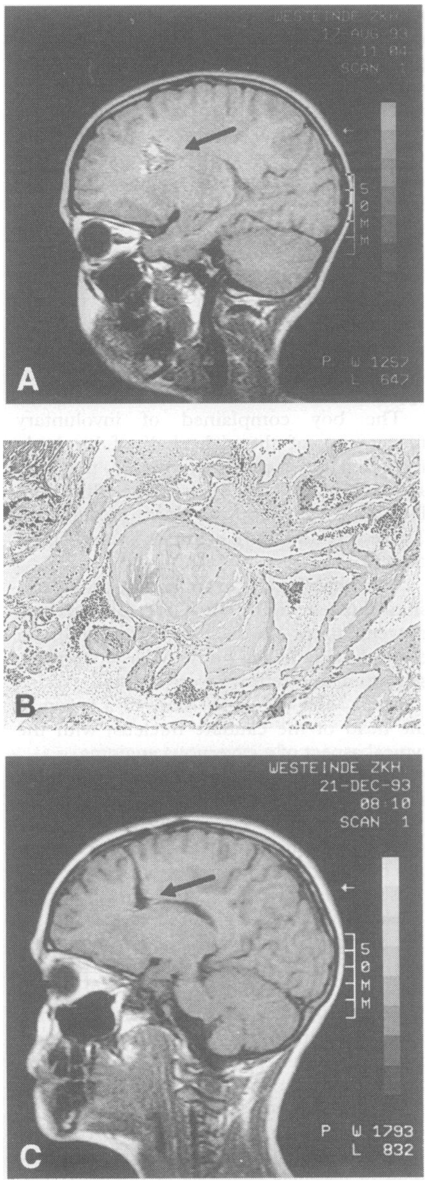
With the Leksell stereotactic frame (Elekta Co, Sweden) the shortest route to the lesion via the paramedial frontal lobe was estimated. At that spot a small burr hole was made and a silastic tube was passed to the border of the lesion with a Backlund catheter implantation set. After craniotomy the lesion was reached with the catheter as a guide. The mulberry like vascular lesion was removed completely, including two small haemorrhages.

Histology (figure B) showed a conglomerate of vascular channels of different size. The wall of these channels consisted of a single inner layer of endothelial cells and an outer layer of collagen of varying thickness. Some vascular spaces were occluded by a recent or an organised thrombus and some vessel walls were partly calcified. Iron pigment was found in and around several vessels, as evidence of prior bleeding. The surrounding brain tissue showed pronounced gliosis and deposition of iron.

In the two months after the operation the hemichorea-hemiballism disappeared completely. Control MRI (figure C) showed complete removal of the angioma.

This case is to our knowledge the first in the literature of a histologically confirmed cavernous angioma presenting with hemichorea. Hemichorea has been described in lesions of the caudate nucleus, and is thought to reflect release phenomena caused by a lesion of the striatal neurons projecting to the external globus pallidus.

The incidence of cavernous angiomas remains obscure. In a consecutive series of 11 children operated on for cerebral vascular malformations five were diagnosed to have cavernous angiomas.[3] Scott *et al*[4] state that in some paediatric institutions cavernous angiomas are the most common cerebrovascular malformations encountered. Most cavernous angiomas, however,



(A) Typical aspect of cavernous angioma in the head of the caudate nucleus (MRI image, SE TR 520/TE 25, after intravenous contrast). (B) Cavernous angioma with anastomosing vascular channels, with fibrotic walls and organised thrombus (centre). There is no pre-existing brain tissue between the vessels (haematoxylin-eosin, originally × 100). (C) Control MRI after removal of the cavernous angioma, showing a postoperative defect.

become symptomatic between the ages of 20 and 40 years.

The natural history of sporadic cavernous angiomas is not well known. Two retrospective review studies of presumed cavernous angiomas, detected by MRI, showed that the most frequent presenting symptoms were seizure with 1·5% per person/year exposure, and haemorrhage, with estimated annual bleeding risks of 0·8%, and 0·25% per person/year respectively.[5 6] The risk of recurrent appreciable bleeding in CVMs that presented with haemorrhage is generally suggested to be larger, perhaps comparable with that in classic arteriovenous malformations, and may prove fatal. In children Scott *et al* recommend surgery if the lesion is safely accessible, is currently symptomatic, or shows evidence of having bled in the past.[4]

Results of operation in previously reported cases of CVMs in the basal ganglia presenting with (progressive) hemiparesis, however, have generally been poor.[2] Stereotactic localisation with CT guidance and microsurgical techniques have facilitated surgery in deep paraventricular or basal ganglia lesions, offering now a better perspective in these cases. This is the second case report describing effective surgical management in resolving a movement disorder due to a cavernous angioma of the basal ganglia, without complications. This indication for removal of a CVM should be considered in subsequent cases.

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MATTERS ARISING

The nature of apraxia in corticobasal degeneration

We read with interest the report by Leiguarda *et al*[1] about the nature of apraxia in corticobasal degeneration. The authors concluded that ideomotor apraxia is the most frequent type of apraxia in corticobasal degeneration. We disagree with them about the conclusion, and would like to comment on apraxia in corticobasal degeneration.

We have experienced four patients with corticobasal degeneration, two of whom were reported elsewhere.[2] These four patients presented consistently with asymmetric limb-kinetic apraxia, but with neither ideomotor apraxia nor ideational apraxia. Unlike ideomotor apraxia and ideational apraxia limb-kinetic apraxia is defined as a breakdown of previously skillful movements, manifested by difficulty in making fine finger movements.[3 4] These four patients also had difficulty in making gestures and using objects on the side of the greater clumsiness. Such apraxic disorders could not be considered ideomotor apraxia or ideational apraxia, because limb-kinetic apraxia can cause clumsiness in all praxic acts on the side contralateral to the lesion.[4]

The discrepancy between the report of Leiguarda *et al* and ours may partly be due to the heterogeneity of corticobasal degeneration or varieties of duration of the illness. On the other hand, the following possibilities may account for the differences. Firstly, as mentioned, limb-kinetic apraxia might induce a disorder of symbolic action, which mimicked ideomotor apraxia, leading to the authors' conclusion. Secondly, limb-kinetic apraxia and ideomotor apraxia might coexist. Limb-kinetic apraxia usually occurs on the side contralateral to the lesion, whereas ideomotor apraxia occurs bilaterally. Thus it is possible, as reported by Leiguarda *et al*, that only ideomotor apraxia is detectable on the side of least clumsiness.

Regarding the underlying mechanism of apraxia in corticobasal degeneration, Leiguarda *et al* attributed ideomotor apraxia to dysfunction of the supplementary motor area. However, the role of the supplementary motor area in motor acts still remains controversial. The supplementary motor area may play an important part, as well as the motor cortex, in execution of complex finger movements and may not work as a supramotor centre.[5] It seems likely that the apraxic disorders arise from another cortical lesion. Neuropathological studies have shown that the sensorimotor cortex is predominantly involved in corticobasal degeneration.[6 7] With SPECT, we showed that cerebral blood flow was mainly decreased in the unilateral perirolandic cortices in all four patients. The perirolandic cortical hypoperfusion could account for contralateral limb-kinetic apraxia, as a lesion in the sensorimotor cortex induces limb-kinetic apraxia on the contralateral side.[4] We therefore consider that limb-kinetic apraxia is the most frequent type of apraxia in corticobasal degeneration, even if ideomotor apraxia or ideational apraxia may exist.

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