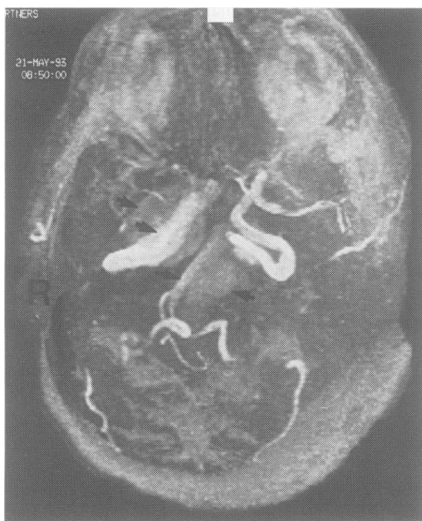


Summary of the literature concerning intracranial fusiform aneurysms

Clinical details	Site of aneurysm	Treatment	Reference
19 year old man. Right retro-orbital headache and diplopia (VI th nerve palsy) Symptoms developed over night	Petrous apex near the cavernous sinus	Selverstone clamp of right internal carotid artery with gradual occlusion	Muhoen <i>et al</i> ²
36 year old man. Right temporal headaches of 3 months duration	Saccular aneurysm and multiple fusiform aneurysms at right middle carotid artery trifurcation	Aneurysmal clip of saccular aneurysm and coating of fusiform aneurysms with methyl methacrylate	Muhoen <i>et al</i> ²
18 year old woman Asymptomatic	Right petrous carotid artery	Nil	Frank <i>et al</i> ³
27 year old man Left sided weakness (arm and leg); left hemisensory disturbance (arm, leg, and face); left sided incoordination and left superior rectus weakness. Symptoms of 1 year duration	Right petrous and cavernous internal carotid artery Basilar artery	Aspirin and β -blocker	



MRI angiogram. Two intracranial fusiform aneurysms are demonstrated: of the internal carotid on the right and of the basilar artery in the centre. In each case, one arrow points to blood flowing through the aneurysm and the other arrow to the region of slow flow or clot within the aneurysm.

patient. His pupillary inequality might be due to disordered autonomic tone resulting from damage to the sympathetic fibres that accompany the intracranial vessels.

Limited CT with contrast showed a large aneurysm of the basilar artery. An MRI scan showed this lesion as well as a second large fusiform aneurysm of the right internal carotid artery as it passed from the petrous bone to the cavernous sinus. Digital subtraction venous angiography and MRI angiography confirmed the presence of these aneurysms and showed a third lesion, a fusiform aneurysm of the extracranial portion of the left internal carotid artery (figure).

This patient thus had three large cerebral aneurysms involving all the major vessels supplying the intracranial contents. As a result these lesions were not amenable to operation. The patient was considered to be at risk of thrombosis and distal embolisation because of slow flow through segments of the aneurysms. Treatment with aspirin was therefore started. β Blockers are recommended for patients with Marfan's syndrome with intracranial aneurysms, the rationale being the reduction of endothelial

wall stress. In view of the limited therapeutic options available for this patient, we considered it prudent to offer him a β blocker even though its benefit is not proved in this context.

The association between neurofibromatosis type 1 and vascular disease is well recognised. Intimal smooth muscle cell proliferation and disordered vascular extracellular matrix, as a manifestation of a generalised mesodermal dysplasia, are possible mechanisms responsible for occlusive and aneurysmal vascular lesions respectively.

Whereas renal and gastrointestinal vascular lesions are common, disordered cervico-cerebral vasculature is less often described. There have been case reports of extracranial occlusive and aneurysmal disease, as well as of intracranial occlusive disease, the second occasionally producing the angiographic appearance of moyamoya disease.¹ Intracranial aneurysmal lesions, however, are very uncommon and, to the best of our knowledge, there have been only three descriptions of (large) intracranial fusiform aneurysm formation in patients with neurofibromatosis type 1 (table).^{2,3} Although histology is unavailable in all of the recognised cases, this description of two intracranial fusiform aneurysms helps to establish the association between this form of angiopathy and peripheral neurofibromatosis.

I am indebted to Dr Mike Wright of the neuroradiology department at Groote Schuur Hospital for his time and assistance in the interpretation of the neuroradiological investigations performed on this patient.

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Mills' syndrome: ascending (or descending) progressive hemiplegia: a hemiplegic form of primary lateral sclerosis?

We describe two patients with slowly progressive hemiplegia. These two cases bring to mind a rare clinical syndrome described in 1900 by Mills—namely, ascending (or, less often, descending) progressive hemiplegia.¹ Mills¹ claimed that this disorder was a new form of degenerative disease characterised by progressive degeneration of the corticospinal pyramidal pathways. Despite its age, the concept of Mills' syndrome is still controversial. Indeed, a number of pathological factors can cause such clinical findings, and the cases that remain isolated and can be considered as primary are rare.

Case 1, a 49 year old right handed woman with no previous personal or family medical history, complained in 1975 of motor deficiency on the right side of the body. The initial symptoms were weakness of the right foot and leg, which slowly progressed to the thigh. The patient was admitted to hospital in 1981 at the age of 55. Physical examination showed right Babinski's and Hoffmann's signs. Tendon reflexes were very pronounced on the right side. The motor deficiency was strictly limited to the right lower limb. There was no sensory loss. An EEG, CSF examination (cytochemistry), EMG, brain CT, and contrast myelography were normal. Re-examination in 1987 showed that the disability had increased: there was a pyramidal gait and a distal motor deficit of the right arm associated with a moderate hypertonia. Facial mobility was normal. No sensory deficit was noted. Routine laboratory tests were normal. Serological tests for syphilis were negative. Examination of CSF showed a slight increase in protein content (0.60 g/l) without pleocytosis; immunological tests did not disclose intrathecal synthesis of immunoglobulin or oligoclonal IgG bands. Visual, auditory, and somatosensory evoked potentials were normal. EMG was performed again with normal findings. MRI of the brain and spinal cord did not show any lesions. Spinal angiography showed no evidence of vascular abnormality. We have not had the opportunity of re-examining this patient since 1987 but the referring physician has kept us informed. In April 1993 after 18 years of evolution he noted a persistent right hemiparesis with hyper-reflexia, Babinski's and Hoffmann's signs, and ankle clonus; there was urinary urgency but no sensory loss, fasciculations, or amyotrophy. Brain and spinal cord MRI and a further EMG were still normal.

Case 2, a 25 year old right handed female nurse first noticed weakness in her left hand in 1976. In ensuing years the weakness progressively spread to the whole arm. In 1985 weakness appeared in her left leg and she began to have trouble in walking. The patient had no personal or family history of neurological disorders. In 1989 physical examination revealed a global weakness of the left upper and lower extremities with pyramidal gait and no sensory deficit. Tendon reflexes were increased in all four limbs with Babinski's and Hoffmann's signs on the left. A facial asymmetry was noted on her left side when the patient was asked to make a face. Mild wasting without fasciculation was noted in the left leg. Laboratory findings, including assessment of the

immune system, detection of inflammation, and serology testing for syphilis were normal. An EEG and visual, auditory, and somatosensory evoked potentials were normal. Brain and spinal cord MRI was normal. Analysis of CSF gave a protein content of 0.10 g/l with no pleocytosis and normal immunological findings. Three EMGs performed at two-year intervals (1985, 1987, and 1989) showed no evidence of denervation. A muscle biopsy performed in 1985 was normal. In June 1993, after 17 years of evolution, the patient's condition was stationary.

Mills described eight cases of a very slow advancing form of hemiplegia beginning usually in the extremity of a lower limb, then spreading up to the homolateral upper limb.¹⁻³ Five more cases were published between 1927 and 1951.⁴⁻⁷ In those 13 and in our two cases pyramidal signs were always seen on the side of hemiplegia and often bilaterally (seven of 15 cases). A moderate amyotrophy without fasciculations is common (six of 15). The palsy can involve the face (five of 15). Sensory disturbances are usually absent but in a few cases (three of 15) minimal hypaesthesias have been noted. The manifestations very gradually worsen. Progression is more often ascending (13 of 15) than descending (two of 15). Involvement of both sides of the body has been reported in advanced stages (five of 15). A family history of the syndrome was not noted in any of the cases. The clinical picture presented by our two patients is identical to that described by Mills.¹⁻³ After an 18 and 17 year duration of development all manifestations were still unilateral except for increased tendon reflexes in case 2.

Mills' syndrome is supposedly due to primary degeneration of the corticospinal pyramidal pathways. Pathological examination has been performed in only one case: the results showed non-specific, staged, irregular lesions of the pyramidal tracts predominating on one side at the level of the spinal cord and in the brainstem. The motor cortex was not involved; Betz cells were normal.

The scarcity of case reports and the availability of only one case confirmed by necropsy raise doubt as to the authenticity of this syndrome. Moreover, many reports date back to times when diagnostic methods did not allow reliable differential diagnosis. In this regard it should be said that ascending hemiplegia with Mills' syndrome-like features could be due to a variety of causes—for example, brain tumours, abnormalities of the cervical spine or cervico-occipital junction, small lacunar infarctions, pontine lesions, multiple sclerosis, or early stage amyotrophic lateral sclerosis. In our two cases the very slow progression of symptoms and the absence of any peripheral motor neuron involvement are inconsistent with amyotrophic lateral sclerosis. Similarly brain MRI, CSF examination, and evoked potentials in both cases ruled out multiple sclerosis. Finally, compression of the cervical spinal cord was not found.

Mills' syndrome could be considered as a variant of primary lateral sclerosis, which is characterised by a pure upper motor neuron syndrome related to a bilateral degeneration of the corticospinal pyramidal tracts. Several cases of primary lateral sclerosis have been documented after elimination of

all other causes of spastic syndromes.⁸⁻¹⁰ Pringle *et al*¹⁰ proposed clinical and laboratory diagnostic criteria for primary lateral sclerosis: all of them are present in our two cases except for negative Lyme disease and HTLV-1 serologies which have not been tested. But Pringle *et al*¹⁰ specified that these tests have to be done in endemic areas only and the south east of France is not such an area. Thus we propose that Mills' syndrome could be considered as the hemiplegic form of primary lateral sclerosis. We hope that this report will incite others to publish similar findings. Many more cases documented with current diagnostic technology will allow the confirmation or otherwise of the nosological authenticity of Mills' syndrome.

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Separation of voluntary and limbic activation of facial and respiratory muscles in ventral pontine infarction

Selective paralysis of voluntary as opposed to limbically activated respiration has been described in a lesion affecting the ventral pons. We report a case in which we have also observed the activation of separate pathways serving voluntary and emotionally influenced facial expression as well as respiration.

A 27 year old white man developed sudden neck pain spreading to the right side of the face associated with deafness in the right ear and profuse vomiting. Two hours later he developed difficulty in walking, sonorous breathing, and episodes of shaking of all four limbs without loss of consciousness.

He was unable to communicate and held his teeth clenched. On admission to hospital he developed a transient (10 minute) left hemiplegia followed three hours later by a complete right hemiplegia. Two hours later he became tetraplegic. Subsequent examination showed that he was responsive and alert: eye movements were full, volitional movements of the jaw, face, palate and tongue were absent but spontaneous and volitional blinking was preserved. He was unable to cough or take and hold his breath to command but spontaneous respiratory rhythm was preserved: he tended to obstruct his upper airway but when this was alleviated by intubation and, later, a tracheostomy, ventilation was adequate. He had an increase in extensor limb tone and the tendon reflexes were brisk with extensor plantar responses. Sensory testing was normal. General examination was unremarkable.

Magnetic resonance imaging showed an area of infarction in the right side of the pons extending inferiorly across the midline (figs 1 and 2). Cerebral angiography showed filling of the posterior inferior cerebellar arteries bilaterally but no contrast entered the basilar artery supporting the diagnosis of occlusion of the basilar artery. Clotting studies, other blood investigations, and echocardiography were normal.

Five days after the onset of the illness the patient was self ventilating via a tracheostomy when awake or asleep, with normal oxygen saturations and PaCO₂. The respira-

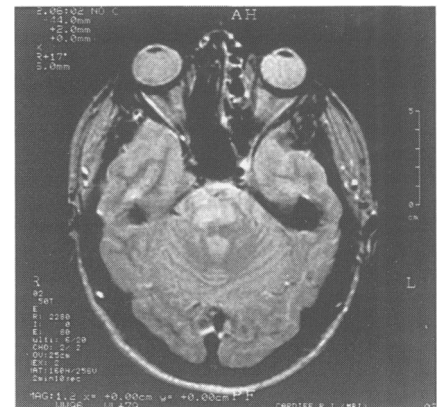


Figure 1 T2 weighted axial MRI at the level of the pons showing an area of increased signal in the ventral aspect of the pons extending across the midline to the left.



Figure 2 T1 weighted midline sagittal MRI showing a low signal area in the ventral pons.