medial infarctions is known as hemimedullary (Babinski-Nageotte) syndrome.⁵ However, because of the separate arterial topography supplying the medulla, the simultaneous occurrence of ischaemic lesions of its medial and lateral parts would be extremely rare.^{6,7} We report MRI of a patient with an ipsilateral lateral medullary infarction (Wallenberg's syndrome) followed by a medial medullary infarction with a 6 month interval. The initial stroke event on the central facial weakness of this patient was previously reported.⁸

A 40 year old man noted weakness of the right limbs and dysarthria. Since the age of 38, the patient had been on an antihypertensive drug regimen. He was conscious, and his blood pressure was 134/80 mm Hg. Neurologically, there were noted right supranuclear facial weakness, paralysis of the left hypoglossal nerve, paralytic dysarthria, and right hemiparesis with prominent weakness of the arm. Deep tendon reflexes were exaggerated in the right limbs and Babinski's sign was positive on the right side. Light touch and vibratory sensations were slightly decreased in the right leg, but temperature and pain sensations were well preserved. There was no diabetes mellitus or hyperlipidaemia. No abnormalities were noted on ECG, transthoracic echocardiogram, and Holter monitoring. Brain MRI at 10 days after the onset showed a high signal intensity area (T2 weighted image) in the left ventromedial part of the upper medulla, which was diagnosed as an infarct (figure A). Cerebral angiography performed at 12 days after onset showed mild atherosclerotic changes of the vertebralbasilar artery. The right facial weakness and left tongue paralysis were gradually improved and the patient was discharged with persisting mild right hemiparesis after one month.

Six months later, however, he suddenly experienced a floating sensation and gait difficulty. At that time he was alert and had a blood pressure of 160/106 mm Hg.

Axial T2 weighted MRI (5000/96, TR/TE). High signal intensity areas (arrows) are noted in the left ventromedial part of the upper medulla (A) and the left dorsolateral part of the upper to middle medulla (B) 6 months after the first stroke.

Neurological examination showed left Horner's syndrome, skew deviation with slight downward position of the left eyeball, clockwise rotatory nystagmus, impairment of temperature and pain sensations of the left face, paralysis of the left soft palate, hoarseness, slight dysphagia, cerebellar ataxia of the left limbs, decreased temperature and pain sensations of the neck and all parts of the right side, and hypohidrosis of the left half of the body. Slight right hemiparesis was present. Brain MRI performed on the next day showed a high signal intensity area (T2 weighted image) in the left dorsolateral portion of the upper to middle medulla. A repeat MRI study at one month after this second attack clearly showed an infarct in a left dorsolateral portion of the medulla with a reduction of oedema, and showed a reduction of an infarct in the left ventromedial part from the first stroke (figure B).

The patient presented here developed a left dorsolateral medullary infarction 6 months after the onset of an infarction involving the left ventromedial part of the upper medulla. Both were clearly seen on MRI. The dorsolateral part and medial part of the medulla are supplied by separate branches from the posterior inferior cerebellar artery and the anterior spinal artery, both of which are indirectly supplied by the vertebral artery. Hence the occlusion of the vertebral artery could produce simultaneous hemimedullary syndrome.^{2,6} However, serial infarctions of the separate topographic areas of the medulla resulting in a hemimedullary syndrome have been suggested,^{3,5} serial events documented by MRI—such as in the present patient—have not been reported. The present case provided unequivocal evidence that the hemimedullary syndrome can occur as serial events of the medial and lateral medullary infarctions.

With the advent of MRI, it has recently become possible to see microvascular lesions in the brain stem that would in the past have been detected only by postmortem examination. In patients where such multiple ischaemic lesions occur at separate sites and times so as to form a certain distinct syndrome, serial observations with MRI will certainly prove the pathogenic background of this syndrome and be of great clinical interest.

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Oculo-auriculo-vertebral spectrum disorder (Goldenhar "syndrome") coexisting with schizophreniform disorder

Oculo-auriculo-vertebral spectrum disorder is a complex and heterogeneous inborn condition affecting primarily aural, oral, and mandibular development with an estimated incidence of between 1/3500 and 1/26 000 births. It is thought to be connected to the development of the first and second branchial arch. It may additionally affect the skeletal system, kidneys, heart, gastrointestinal tract, and eye. Familial occurrence has been reported, but most cases seem to be sporadic.¹ Structural CNS and neurological abnormalities occur often, but with great variability.² Mental retardation seems to be a common feature,¹ but other relevant psychiatric disorders have not been reported.

We present what seems to be the first published case of a patient with an organic schizophreniform disorder and oculo-auriculo-vertebral spectrum disorder.

A 27 year old man was referred for psychiatric treatment by his general practitioner. The patient reported intermittent speech problems since the age of 9 which, he was unshakably convinced, were the effect of "electromog". He would often smell strange and sickening odours originating in his mother's kitchen that nobody else would notice and were causing his problems. Officially he was studying business administration, but had not passed any of the necessary exams in the past 3 years. Mentally he thought he was absolutely sane.

From birth he showed facial asymmetry, right thumb dysplasia, and dysplasia of the right auricle. Neither the patient nor his mother knew of a relevant family history of psychiatric disorders or dysplasias of any kind. There were no further psychiatric treatments except for one short admission for somatisation disorder 2 years previously. No prenatal or perinatal abnormalities were known. The patient's school performance had been average to good. He had first successfully trained and worked as a chemical laboratory assistant, but he had discontinued a subsequent polytechnic engineering course for no obvious reason.

On psychiatric examination the patient was suspicious and reserved. Thought contents were restricted to his complaints; formal thought showed a loosening of associations and often tended to be illogical. Attention, comprehension, and concentration were mildly impaired. There were olfactory hallucinations (kitchen smells), but no other

sensory impairments. Delusional ideas—but no autochthonous delusions—were present (control by electrosmog), they were not systematised and had a low affective impact. There were no delusions of reference. Mood was blunt and flat. There were no psychomotor abnormalities, but marked apathy was noted. Neuropsychological testing showed normal intelligence, normal visual memory, reduced speed of visual processing, and, despite the discrete attention deficit, normal accuracy, concentration, and speed. Physical examination confirmed dysplasias of the right auricle and the right thumb as well as scoliosis of the cervical and thoracic spine. Cranial nerves were unaffected, but right sided conductive hearing loss (audiometrically confirmed) and marked facial asymmetry with a hypoplastic right half were prominent. Ear, nose, and throat examination showed submucous cleft palate, malocclusion and hypoplasia of the maxilla, mandibular asymmetry, and cluttering with mild stuttering.

Routine blood and urine tests were normal. Electrocardiography, echocardiography, and chest radiography showed no cardiac or pulmonary abnormalities. Radiography showed right sided rib deformation, aplasia of the left 12th rib, a hypoplastic atlas, a plump odontoid process, basilar impression, complete (C5/7) and incomplete (C2/3) fusion of vertebrae, rotational scoliosis, and segmental synostoses of the cervical and thoracic spine. Asymmetry of the mandible and hypoplasia of the maxilla was cephalometrically confirmed on skull radiography. Cranial MRI showed a hypoplastic vermis cerebelli inferior but no other abnormalities. An EEG showed no ictal activity. Cortical magnetic stimulation was normal, whereas the blink reflex showed markedly lowered responses on the right side (trigeminal nerve; R1) and the masseter reflex produced no right sided electrophysiological response, but normal responses on the left. Acoustic evoked potentials showed a peripheral conductive delay on the right side. The karyotype was 46, XY. Fluorescence in situ hybridisation techniques showed no microdeletions on chromosomes 22q13.3 or 10p13/14.

We treated the patient with the atypical neuroleptic drug olanzapine (15 mg/day) and supportive psychotherapy. This resulted in a mild but significant amelioration of the disturbances of thought, concentration, attention, and comprehension, the rather low intensity olfactory hallucinations and delusional ideas diminished markedly.

This patient with normal intelligence exhibited multiple abnormalities: oral (submucous cleft palate, malocclusion), phoniatric (cluttering with stuttering), auricular (right sided conductive hearing loss and dysplasia of the auricle), facial (asymmetry), skeletal (right sided aplasia of the 12th rib, fused cervical vertebrae, and other dysplasias of the cervical and thoracic spine, abnormalities of the cranial base, right thumb dysplasia). Oculo-auriculo-vertebral spectrum disorder lacks clear minimal diagnostic criteria, but according to Gorlin *et al.*¹ our patient would qualify for this diagnosis.

There is no final answer to the question whether this schizophreniform disorder is idiopathic or symptomatic, as an unequivocal distinction between idiopathic and symptomatic schizophrenia cannot be made by phenomenology.³ But despite the age of the patient, there are arguments for a symptomatic aetiology: The lack of autochthonous (or primary) delusions, of delusions of refer-

ence, and of Schneiderian first rank symptoms, the exclusively olfactory hallucinations, the absence of a positive family history of psychiatric disorders, the neither schizoid nor schizotypal premorbid personality, and the deficient pattern of thought disorders and cognitive abilities and at the same time the organic abnormalities affecting other systems of the head (speech, cleft palate, cranial base, hearing loss, facial asymmetry, cerebellar structures) support the idea of an organic background to the disorder.³ We think that inborn syndromes of the head and neck—particularly oculo-auriculo-vertebral spectrum disorder—have received too little attention from neuropsychiatrists. Given their embryological position, these conditions might help develop some hypotheses about the aetiology of psychiatric disorders.

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Disabling stroke arising five months after internal carotid artery dissection

Dissection of the internal carotid artery is an increasingly recognised cause of acute ischaemic stroke in young adults and children. It may arise spontaneously or secondary to blunt or penetrating trauma. It has been reported after seemingly trivial incidents, such as reversing a car, washing hair, or holding a mobile telephone by flexing the neck against the shoulder. The incidence of carotid dissection is about 2.5–3/100 000/year—similar to aneurysmal subarachnoid haemorrhage.¹ The most common presenting features are ipsilateral temporal, retroorbital, or hemicranial pain, Horner's syndrome, and local cranial nerve palsies, plus potentially devastating cerebral ischaemic events. Although no trial data exist to support the use of anticoagulants, most cerebrovascular specialists advocate initial intravenous heparin then three to six months of warfarin treatment if there have been ischaemic episodes. There is often an interval between onset of symptoms and cerebral infarction enabling the diagnosis to be made and treatment to be instituted.² Given the potentially fatal or disabling consequences of carotid dissection this window of opportunity is not to be missed. About 80% of ischaemic strokes arise

within the first seven days although they can occur four to five weeks after the onset of symptoms.^{2,3} We report a patient who developed a disabling stroke five months after ipsilateral carotid dissection. This has implications for instituting treatment in a patient seen weeks or months after the incident, and for the duration of subsequent anticoagulation.

A 24 year old man was involved in a road traffic accident. Emergency fire services were required to free him from the vehicle and he remains amnesic for the event. He sustained soft tissue injuries to the face, chest, and arms, but no fractures. No surgery was required. No neurological sequelae were noted although he mentioned mild visual blurring in retrospect. A non-contrast CT was performed the day after the accident which showed a small right peripheral parietooccipital infarct (but this had been reported as a contusion). The patient was discharged and made a full recovery from his injuries.

About four months later he suddenly developed a left facial weakness. There was associated right retro-orbital and temporal headache. Over three to four days the headache and facial weakness cleared. Medical attention was not sought.

Just over five months (158 days) after the traffic accident he was admitted to our unit. That morning he had suddenly developed a dense left hemiparesis affecting the arm and face more than the leg. The right sided headache had returned. A repeat CT showed maturation of the previous occipitoparietal infarct and a new infarct in the corona radiata on the right, extending into the internal capsule. A colour flow Doppler/duplex scan of the cervical carotid arteries showed normal appearances on the left side, a normal right external carotid artery, but reduced diastolic flow in the right common carotid artery. The origin of the right internal carotid artery was patent but the Doppler waveform was severely damped with the typical "bidirectional" signal associated with distal occlusive disease and commonly seen in dissections. The diagnosis of dissection of the right internal carotid artery was confirmed by T1 weighted axial MRI of the neck. These showed characteristic high signal in the vessel wall due to haemorrhage with associated reduction in diameter of the true lumen. The patient was treated immediately with intravenous heparin. Seven days later warfarin was started for six months. His hemiparesis gradually improved.

The right internal carotid artery probably dissected at the time of the road traffic accident and the infarct seen on the initial CT had the characteristics of a peripheral embolic lesion in the territory of posterior superficial middle cerebral artery branches. A second (presumably) embolic insult causing transient facial weakness arose four months later. Another five weeks passed before the disabling left hemiparesis. During the five months since the traffic accident neither the patient nor his family could recall any occasions when he had complained of a new neck or head pain relating to further neck trauma. It seems likely that he had had only one insult to the right internal carotid artery but that over five months the vessel had failed to heal spontaneously and the damaged intima continued to act as a focus for thrombus formation.

Recurrent dissection of the same extracranial vessel is extremely unusual, and occurred in only one of 81 patients followed up for a