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 reference, 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

median of almost three years by Bassetti *et al*.[4] Recurrent dissection involving other extracranial vessels is slightly more common, with an incidence of about 1% a year.[5] Our patient had no clinical evidence of the heritable connective tissue disorders sometimes associated with multiple or recurrent dissections (Marfan's syndrome, Ehlers Danlos type IV, etc). He clearly had three ischaemic episodes over five months attributable to the right internal carotid artery and we think that persisting damage, rather than true recurrence, was the likeliest cause of his stroke.

It is not uncommon to be asked to see young patients with ischaemic stroke several weeks after the event. Even when arterial dissection is suspected or proved, if the patient has had no further episodes for a month or more it is tempting to prescribe antiplatelet agents rather than submitting the patient for formal anticoagulation. This case shows that disabling stroke can occur as long as five months after the initial dissection; thus anticoagulation should probably be considered even when there is considerable delay in referral. Although empirical, a minimum period of six months on warfarin would seem appropriate. If repeat duplex ultrasound remains abnormal at that time, extension to 12 months may be necessary.

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Severe weight loss after withdrawal of chronic pizotifen treatment

A 36 year old woman was admitted to hospital for investigation of weight loss of 10 kg over 2 months. She had had classic migraine for over 20 years, and had been taking 1 mg pizotifen (Sanomigran) daily continuously as migraine prophylaxis with good effect. Eight weeks before admission, this drug had been discontinued. A week later, she developed frontal headaches with nausea. She became anorexic and began to lose weight quickly. General and full neurological examination showed no abnormality. Investigations including full blood count, erythrocyte sedimentation rate, urea and electrolytes, liver function tests, random cortisol, blood glucose, and thyroid function tests were all normal. A pregnancy test was negative, and gastroscopy, including duodenal biopsy showed no abnormality. Brain CT was normal. Pizotifen was restarted and she immediately felt better. Her appetite improved, headaches stopped, at 1 month she had regained 5 kg, and by 2 months was back to her normal weight, remaining asymptomatic.

Pizotifen is widely used for migraine prophylaxis, where it modifies humoral mechanisms inducing headache by effects on serotonin and histamine.[1] Treatment is often associated with increased appetite, a craving for carbohydrates, and weight gain, probably induced by its powerful antiserotonin activity, an effect which has been used clinically in the treatment of anorexic and convalescent patients.[2] Although weight loss could theoretically follow discontinuation of chronic pizotifen treatment, this does not seem to be a noted side effect, and there are no published reports of severe weight changes. However, the manufacturers (Sandoz Pharmaceuticals) received a similar case report to this in 1986 in which a patient who had taken 1.5 mg daily for 2 years had lost 2 stone over 2 months after withdrawal of the treatment. Hence, it seems that marked body weight reduction may follow withdrawal of long term pizotifen treatment, and knowledge of this adverse effect may prevent extensive and unnecessary investigation in those subsequently presenting with anorexia and severe weight loss.

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Successful treatment of intractable epilepsia partialis continua with multiple subpial transections

Cortical dysplasia is increasingly being recognised as an important cause of partial seizures including epilepsia partialis continua. With the advent of high resolution MRI it is now often possible to identify areas of cortical dysplasia, increasing the possibility of neurosurgical intervention when seizures are refractory to medical treatment. We present a patient with intractable epilepsia partialis continua due to cortical dysplasia that was refractory to all medical treatments, was not evident on MRI, and was dramatically improved by multiple subpial transections.

A 19 year old man who was the product of an uneventful pregnancy with no perinatal problems and normal early milestones developed simple partial seizures at the age of 14 years. These comprised clonic movements of the left side of his face, left arm, and left leg. Brain MRI was reported as normal and an EEG showed a right frontoparietal focus. Carbamazepine was started at a dose of 800 mg daily and he remained seizure free for a year. A subsequent recurrence of the left sided simple partial seizures responded to the addition of phenytoin (300 mg daily). Over the next four years there was reduced academic attainment but seizures were infrequent. At the age of 19 he developed clonic movements of the right side of his face and right arm. These partial seizures increased in frequency and after a week he was admitted to his local hospital with epilepsia partialis continua comprising continuous motor seizures of the right side of his face and right arm. There was no alteration of consciousness ictally but his seizures were sufficiently distressing to require sedation and ventilation. Several antiepileptic drugs were tried over a two month period without success, including phenytoin, phenobarbitone, sodium valproate, vigabatrin, and infusions of clonazepam, lorazepam, and diazepam. The patient was transferred to this hospital, ventilated, and sedated on a thiopentone infusion. On examination there was marked hepatosplenomegaly but no focal neurological deficit was evident. Investigations were aimed at identifying a progressive degenerative disorder in view of the cognitive decline, involvement of both hemispheres at different times, and the hepatosplenomegaly. A full metabolic screen was normal. Liver, bone marrow, skin, and muscle biopsies were noncontributory and a screen for mitochondrial DNA mutations was negative. The MRI was performed on a 0.5 T Vectra (GE) instrument with T1 and T2 weighted axial and coronal sequences including volume acquisitions, and again no abnormality was detected. His EEG showed spike and sharp waves over the left central cortex, and during seizures runs of repetitive polyphasic discharges were recorded. Electrocorticography and a biopsy of the left premotor cortex was performed. The cortex appeared macroscopically normal but electrocorticography showed polyphasic discharges over the area that was biopsied. Histological examination showed areas of focal cortical dysplasia with disordered cortical lamination, large dysplastic neurons occurring in clusters and aggregates of astrocytes (figure). During this period of investigation his seizures could only be controlled initially on a thiopentone infusion at doses sufficient to cause burst suppression on EEG. This resulted in impairment of liver function and coagulopathy and so was therefore replaced with a propofol infusion. High doses of up to 1000 mg/hour were required to control his seizures, but this was associated with prolongation of bleeding time and gastrointestinal haemorrhage and had to be discontinued. Other antiepileptic drugs were tried without success. Vigabatrin at a dose of <4g daily, carbamazepine <3g daily, gabapentin <2.4 g daily, piracetam <24 g daily, and acetazolamide <1 g daily did not control seizures. Phenytoin induced myelosuppression and was withdrawn. Some degree of control was obtained with a combination of ketamine, phenobarbitone, and midazolam but at doses that rendered the patient unconscious. In view of the intractable nature of the seizures and biopsy findings, multiple subpial transections were undertaken. Before the transections were performed high amplitude spikes were recorded over the left premotor and motor cortex on electrocorticography. Guided by the electrocorticography, multiple subpial transections were performed on the left precentral and postcentral gyri and after this the electrocorticography showed no epileptiform discharges. Postoperatively there was marked improvement in seizures, it was possible to discontinue intravenous antiepileptic drugs and the patient was extubated. The hepatosplenomegaly resolved. He subsequently had only a few brief self limiting clonic movements of the right arm. There was no neurological deficit attributable to the surgical procedure but rehabilitation was hampered by a critical illness polyneuropathy. Nine months after discharge he remains wheelchair bound but is beginning to mobilise, with no recurrence of his EPC.

Cortical dysplasia is increasingly being recognised as an important and treatable cause