axonal polyneuropathy should include biopsy of the minor salivary glands, even when there are few arguments in favour of the diagnosis of primary Sjögren's syndrome.

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Reliability of clinical diagnosis of Huntington's disease

Huntington's disease has a prevalence of between four and 10 per 100 000 in the United Kingdom.[1] It has severe and progressive physical and psychiatric effects. Onset symptoms are reported to be neurological in 46% of cases, psychiatric in 36%, and combined neurological and psychiatric in the remainder.[2] There is thus considerable potential for misdiagnosis.

The cloning of the gene responsible for Huntington's disease showed that the disorder is caused by an expansion of a CAG trinucleotide repeat in the 5' transcribed region. The original report[3] suggested that normal people had 11 to 34 copies of the repeat and those affected with Huntington's disease had 42 to 100 copies. Our own studies with an improved polymerase chain reaction assay that measures only the specific size of the CAG repeat[4] show that the copy number in normal subjects extends from 8 to 33, whereas the lower end of the Huntington's disease range starts at 35.[5]

As we find no overlap between the two distributions, it is possible to use CAG measurement to estimate how often Huntington's disease is clinically misclassified. We have already reported three incorrect diagnoses in a series of 340 with purported Huntington's disease (0·9%), made up of one with presenile dementia of the Alzheimer type, one with multi-infarct dementia, and one with Parkinson's disease.[5] We have now searched for missed Huntington's disease diagnoses in a series of 221 patients with diagnoses of schizophrenia, 79 with presenile dementia of the Alzheimer type, and 68 with senile dementia.

The figure shows the distributions of CAG repeats. There were two possible missed cases of Huntington's disease in the set of 368 patients with psychiatric disorders. One patient, who died at age 88 after a stay in hospital of 42 years and a diagnosis of schizophrenia, had a CAG repeat size of 36. There was no family history of Huntington's disease. At necropsy the brain was removed and fixed intact for a complete neuropathological study. Findings were consistent with a diagnosis of schizophrenia and no abnormality was detected in the caudate. The second patient, who died at age 68 of presenile dementia of the Alzheimer type, had a CAG repeat size of 34. There were no extrapyramidal signs of Huntington's disease at necropsy. Re-examination of the case notes and a further report from medical and nursing staff caring for the patient suggested no symptoms of Huntington's disease.

In all other respects the CAG distributions among the psychiatric disorders were identical to the distribution among normal subjects. There is currently debate about the existence and extent of possible overlap between the normal and Huntington's disease CAG repeat sizes. Although this is yet to be resolved, our finding of a maximum of two missed cases of Huntington's disease (if that is what they were) in 368 patients with psychiatric disorders should increase confidence in the new molecular assay.

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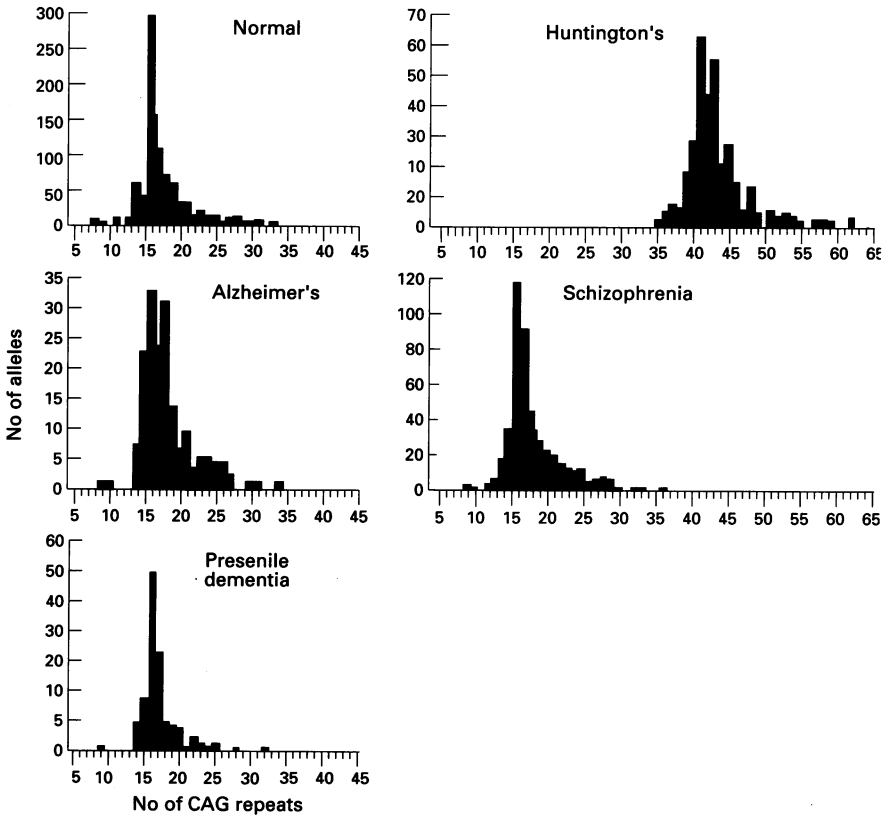

Distribution of CAG repeats in normal subjects and patients with various psychiatric disorders.

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Effect of sudden episodic intracranial hypertension on the electroencephalogram in a child with head injury

There is some controversy regarding the treatment of raised intracranial pressure, particularly in the very young, where the upper limit of normal intracranial pressure is below 5 mm Hg.[1] There is a general consensus that active treatment should be instituted for sustained intracranial pressure of 25 mm Hg or greater in adults. Intervention in children, however, needs to take account of the lower values of intracranial pressure, blood pressure, and cerebral perfusion pressure, and there are limited data for critical thresholds in children. Ideally treatment should be based on multimodality monitoring of cerebral blood flow and metabolic function.[1] This case report shows the functional consequence on brain electrical activity of acute intracranial hypertension and concomitant changes in cerebral perfusion pressure in a young child with head injury.

An 18 month old male child sustained a severe, coma producing, non-accidental

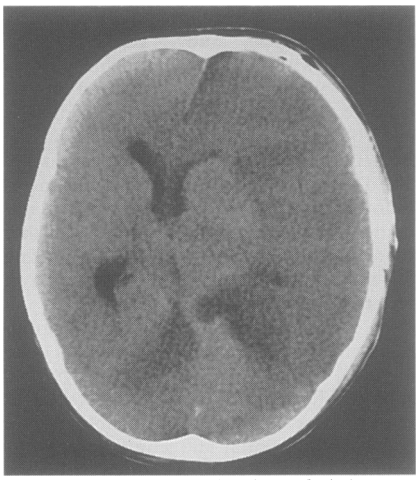

Figure 1 Brain CT of head on admission showing mass effect and midline shift.

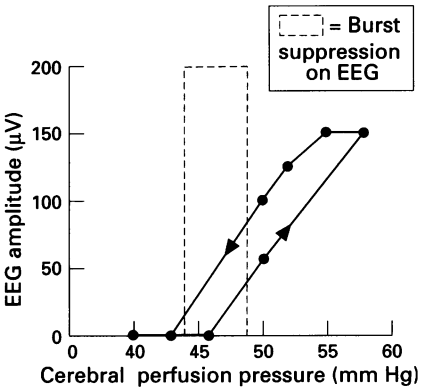

Figure 2 Amplitude of EEG plotted against cerebral perfusion pressure.

head injury. Brain CT showed a left sided acute subdural haematoma with mass effect (fig 1). The haematoma was evacuated promptly and an intraventricular catheter inserted to measure the intracranial pressure. The mean pressure was initially raised and thiopentone was therefore given to maintain it at 20 mm Hg or below. The patient was mechanically ventilated for 21 days, but was weaned off barbiturates after 10 days. During weaning, an eight channel EEG was recorded from Ag/AgCl scalp electrodes placed according to the International 10/20 system with mastoids linked in common reference. After recovery from the barbiturate induced burst suppression pattern, the EEG was dominated by unorganised polymorphic slow activity, which was severely attenuated over the left hemisphere. Transcranial Doppler ultrasound of the middle cerebral arteries showed mean flow velocities of 58 cm/s on the right and 16 cm/s on the left, indicative of hypoperfusion on the left. An increase in intracranial pressure caused a reduction in peak to peak amplitude of the EEG, a further increase produced burst-suppression, and finally electrocerebral silence occurred seconds after the intracranial pressure increased to 30 mm Hg (cerebral perfusion pressure = 45 mm Hg; mean arterial blood pressure − intracranial pressure). In the absence of a mechanical cause, immediate hyperventilation reduced intracranial pressure and as cerebral perfusion pressure exceeded 50 mm Hg the EEG activity returned (fig 2). The child regained consciousness without hemiparesis, but still had neurological impairment one month after his injury.

Intracranial pressure is best interpreted with either a neurological examination or some measure of cerebral blood flow (for example, transcranial Doppler ultrasound) and metabolism (EEG or jugular venous oxygen).[1] This single case study in a young child shows a critical intracranial pressure of 20 mm Hg for cerebral electrical activity. This level marks the transition from a favourable to a poor outcome in severely brain injured adults[2] and children.[3]

It has been suggested that cerebral perfusion pressure is the critical variable that determines outcome in severe brain injury.[4] In adults the optimal level is generally considered to be 60 mm Hg, although recent findings indicate that it should be maintained above 70 mm Hg.[4] A study in head injured children found a critical lower limit of 40 mm Hg, but the authors suggested that the principal aim of treatment should be to achieve a cerebral perfusion pressure of 50 mm Hg or more.[5] The changes in the EEG in this child provide functional evidence for a critical value of 50 mm Hg and therefore support such a protocol.

In children several investigators have reported persistent cerebral blood flow and glucose metabolism in brain-dead children with electrocerebral silence.[6] This dissociation of cerebral blood flow and metabolism from electrical activity has been explained by the expansile skull and fontanelles in young children and the activity of inflammatory microglial cells in the ischaemic cortex. The EEG may therefore be an acceptable alternative to other measures of cerebral blood flow and metabolism for the functional assessment of intracranial pressure, and the rationale for its treatment in young children.

In conclusion, these findings support the need for multimodality monitoring to interpret intracranial pressure measurements,[1 4] and highlight potential advantages of the EEG in children. The aim of treatment in brain injured children should be to maintain intracranial pressure below 20 mm Hg and cerebral perfusion pressure above 50 mm Hg to optimise outcome.

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Unusual presentation of a germ cell neoplasm

According to two reports, a metastasis to the CNS is a common complication of disseminated germ cell tumours of the testes. Metastatic disease occurred in 16% of 242 patients treated in one series[1] and in 15·1% in another.[2] The prognosis for patients with brain metastases from non-seminomatous germ cell tumours of the testes has improved over the past 10 years, as more effective chemotherapeutic protocols have been found.[3]

We document a case in which a patient presented with an intracerebral haemorrhage initially thought to be due to an arteriovenous malformation. A biopsy of the lesion taken at the time of evacuation of the haematoma, however, showed that this was due to an underlying carcinoma.

A previously fit and well 35 year old man presented with a sudden onset of headache associated with nausea and vomiting, while a passenger in his car. On admission he was restless, with a severe headache, but orientated and obeying commands. Neurological examination showed a left hemiplegia. His condition rapidly deteriorated and he showed signs of acute raised intracranial pressure including a dilated, non-reacting, right pupil.

He was initially resuscitated with 20% mannitol and ventilated. A brain CT showed a large right hemispheric intracerebral haemorrhage (fig 1).

An urgent craniotomy was performed. Old and fresh clots were evacuated and there were abnormal arteriolised veins in the periphery of the tumour bed surface

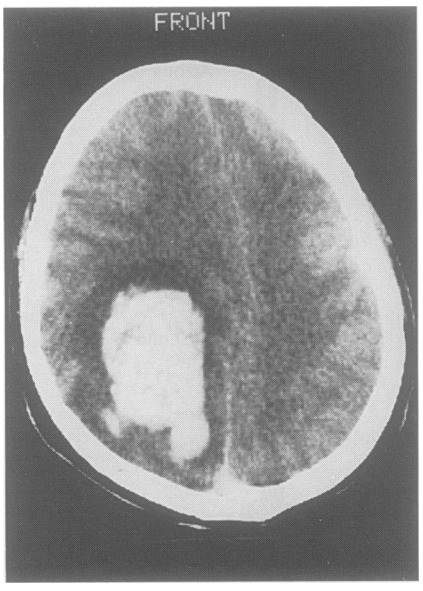

Figure 1 Brain CT showing a large right parieto-occipital haemorrhage.