axonal polynuropathy 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. 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. 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 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 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.

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. We have now searched for misdiagnosed Huntington's disease's 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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Effect of sudden episode 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.2 This case report shows the functional consequence on brain electrical activity of acute intracranial hypertension and concomitant changes in cerebral perfusion in a young child with head injury.

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