that impotence may ensue. The first man encounters no problems; the second develops impotence soon after. A patient satisfaction survey a week later asks both to evaluate my care. The first, free of adverse consequences, praises my approach. The second vilifies me, convinced (possibly correctly) that I induced impotence through my recitation of possibilities. Thus, since their responses may be confounded by both their expectations and outcomes, a summary score of "50% satisfaction" does not convey a helpful message as I try to draw on patient perceptions to improve care.

Where the two patients will probably agree is on what I did. Whatever their expectations or outcomes, both are likely to report that I explained the possible side effects of the new drug. Such reports are far more helpful to those who want to improve care. If I ask patients whether they were told what activities they could or could not do after leaving hospital and learn that a quarter report not being told, I have collected specific, actionable, clinically important data that can inform and shape my future behaviour.[2]

Although the science of learning from patients may be in its infancy, it brings exciting opportunities to clinicians, researchers, and policy makers. For clinicians, data generated from patient reports serve initially as screening rather than diagnostic tests. By analogy, knowing that a screening test for blood in the stool is positive is the first step; determining the cause requires further inquiry. When the Lothian Health Authority finds that a quarter of its patients feel they were not encouraged to ask questions about their treatment,[1] the next step is to search further and find out why. A subsequent series of questions with greater specificity may help in the hunt, just as examining the intestines will help the clinician seeking accurate diagnosis and treatment for blood loss.

For researchers, there is ample and rewarding room for inquiry. They can begin by sharpening questions to patients and their families, learning when and where best to ask them, and developing databases useful to patients, clinicians, scholars, and policy makers. Should one ask for patients' views as they leave a hospital ward or examining room or is it better to wait several weeks and give them time to reflect? How and to what degree do responses differ when gathered face to face or by telephone, by computer or by hand? Do patient reports about the processes of care correlate with clinical outcomes? Questions abound, and the rush of inquiry augurs well for the evolving science.[3-8]

Those who pay for care and those who shape and change the delivery system are also drawing on patient generated data with growing eagerness. National surveys of probability samples of patients provide baseline data and benchmarks, pointing analysts toward both deficient and outstanding practices.[9-11] In the United States consortiums of employers, providers, insurers, and patients are measuring the quality of care in urban and rural settings and are beginning to make major decisions about resource allocation based on their findings.[12] Such assessments of quality now invariably include patient generated data.[13]

At a time when anger and distrust seem ubiquitous in the health systems of so many countries, asking patients to report on the quality of their care may bring clinicians and those they serve closer together. We may break down barriers if we work hard to learn from patients—and then invite them to collaborate in tackling the problems they uncover.

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Diagnosing Creutzfeldt-Jakob disease

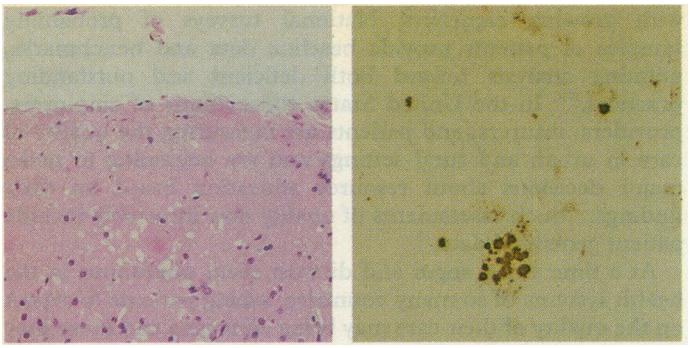
Case identification depends on neurological and neuropathological assessment

Central to the identification and classification of patients with Creutzfeldt-Jakob disease (CJD) is the application of standardised diagnostic criteria based on accumulated data on the clinical and pathological features of the disease. Typically, patients present clinically with rapidly progressive dementia and myoclonus associated with various focal neurological signs. The diagnosis is confirmed by identifying characteristic neuropathological features, which include spongiform change, astrocytic gliosis, and neuronal loss. Diagnostic criteria originally proposed in 1979[1] have been validated by the "gold standard" of experimental transmissibility in primates.[2] This indicates that the clinical diagnosis is highly accurate, particularly if there is a characteristic appearance on an electroencephalogram.

However, not all cases are straightforward: about 10% of patients have a protracted clinical course, which makes the distinction from Alzheimer's disease difficult[3]; 20-40% of patients do not exhibit a typical electroencephalogram[4]; and unusual clinical phenotypes occur in patients with genetic and iatrogenic forms of the disease.[4] Updated diagnostic criteria have been published, including new definitions for "familial" and iatrogenic Creutzfeldt-Jakob disease,[5] but case ascertainment in the small proportion of atypical cases depends on review of a wide spectrum of suspect cases and a high necropsy rate.

Since 1990 in Britain, the criterion for referral to the National Creutzfeldt-Jakob Disease Surveillance Unit has been any suspected case, and about 70% of these cases will go to necropsy. About half of all suspect cases fulfil criteria for definite or probable Creutzfeldt-Jakob disease,[6] reflecting a high level of cooperation from the neurological community in referring any case in which the diagnosis is raised even as a possibility.

Crucial to case ascertainment is the targeting of professional groups who are likely to diagnose patients with Creutzfeldt-Jakob disease. In Britain since 1980, and more recently in other European countries, targeted groups have included neurologists, neurophysiologists, and neuropathologists. Although classic



New variant Creutzfeldt-Jakob disease: (left) haematoxylin and eosin stain shows the characteristic neuropathological feature—amyloid plaque in cerebral cortex (centre) surrounded by a zone of spongiform change; (right) immunocytochemistry for prion protein shows a strong positive reaction (brown) in multiple plaques throughout cerebral cortex

Creutzfeldt-Jakob disease usually presents with rapidly progressive dementia, in about 10% of cases the initial clinical symptom is behavioural disturbance or personality change, often resulting in psychiatric referral. An important assumption in epidemiological surveys is that such patients will be referred for a neurological opinion once neurological deterioration develops. This has been borne out in the British study[7] and also in a study in France.[8]

There has always been concern that elderly patients might be missed, and the significant increase in the incidence of Creutzfeldt-Jakob disease in Britain since 1990[6] is largely due to an increase in the number of cases in patients aged over 75, probably reflecting better case ascertainment in this group. On the other hand, death certificates mentioning Creutzfeldt-Jakob disease have been used in Britain as a safety net for case identification; and since 1990, two thirds of certified cases have fulfilled the diagnostic criteria for Creutzfeldt-Jakob disease, of which only a small minority had not been seen by a neurologist.[4] In Germany, where there is a tradition of joint training in neurology and psychiatry, 1436 departments of neurology, psychiatry, and rehabilitation have been asked to notify cases of Creutzfeldt-Jakob disease. However, the overall incidence in Germany is similar to that in Britain,[9] where clinical referral has largely depended on targeting about 400 neurologists.

The identification of a new variant of Creutzfeldt-Jakob disease in Britain[10] has been possible only through the cooperation of neurologists and neuropathologists and the availability of comparative information from parallel surveillance projects in Europe, coordinated through the European Community's BIOMED 1 research programme. Psychiatric symptoms are a component of the early clinical phenotype of the new variant disease, and in a recent survey only three out of 10 psychiatrists in one British city were aware of the surveillance project.[11] This is hardly surprising as psychiatrists have not been asked to notify cases of Creutzfeldt-Jakob disease in Britain, but it is pertinent to consider whether the system of case ascertainment should be extended to psychiatrists. Review of psychiatric histories in patients with the new variant disease suggests that there is no specific psychiatric phenotype and that distinction from common psychiatric diagnoses such as depression may be impossible.

In all the cases of new variant disease, the psychiatric symptoms were superseded after a period of some months by progressive and devastating neurological dysfunction—including ataxia, cognitive impairment, and involuntary movements—terminating in severe neurological dysfunction and death. Although 11 out of the 12 cases were seen by a psychiatrist, only six patients were initially admitted to hospital under the care of a psychiatrist, and these were promptly referred to a neurologist when the neurological syndrome developed. On current evidence a clinical diagnosis of new variant Creutzfeldt-Jakob disease cannot be made during the psychiatric phase of the illness, and identifying these cases depends on the evolution of neurological signs and in particular on neuropathological examination.

Early diagnosis of new variant disease was made in three cases by brain biopsy, which showed numerous cortical plaques surrounded by spongiform change. However, it is difficult to recommend biopsy as a routine diagnostic tool in view of the risk of complications (such as extradural haematoma or brain abscess), the possibility of sampling an area of brain unaffected by the pathological process, the need to destroy the neurosurgical instruments, and the low chances of the result altering management of the patient. There is an urgent need for an early non-surgical diagnostic test.

Last week saw the publication of details of a new test for Creutzfeldt-Jakob disease, based on immunoassay of 14-3-3 protein in the cerebrospinal fluid, which promises high sensitivity and specificity.[12] The test would be of particular value if it allowed accurate diagnosis at an early stage in the clinical course and in atypical cases of the disease, but this remains to be established."Blind" testing of samples of cerebrospinal fluid from British patients with classic and new variant Creutzfeldt-Jakob disease, done in collaboration with research groups in the United States, has provided promising results, but a specific test for new variant disease is not yet available. Until this is possible the diagnosis of new variant disease will depend on clinical assessment and in particular on the evolution of neurological signs. Confirmation depends on the apparently characteristic neuropathological appearances.

The crucial issue of whether new variant disease is causally linked to bovine spongiform encephalopathy (BSE) may be answered only by continuing epidemiological research, including comparative studies of the incidence in countries with different incidences of bovine spongiform encephalopathy. The methodologies of these studies must be adapted to improve case recognition of new variant Creutzfeldt-Jakob disease, but surveillance will become unmanageable if reference centres are flooded with referrals of patients who turn out not to have the disease. Because new variant disease occurs in younger patients, including teenagers, it is essential to consider extending surveillance to the paediatric population, and lessons drawn from surveillance of classic Creutzfeldt-Jakob disease in elderly people suggest that we should also be alert for new variant disease in this age group. The methods of Britain's Creutzfeldt-Jakob disease surveillance system must adapt to changing circumstances, but case identification still depends primarily on neurological and neuropathological assessment.

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