degree of conformity on medical practice yet allow some forms of manifestly unsatisfactory performance to fall outside the scope of the procedures.

There is thus a strong argument for a broad definition of seriously deficient professional performance rooted in the concept of peer review, on which professional self regulation depends. This would parallel the definition of serious professional misconduct, which in law amounts to that which a responsible body of medical opinion considers to be serious professional misconduct. Seriously deficient professional performance might thus be considered to be a pattern of performance so deficient that, in the opinion of a responsible body of medical opinion, it places patients at risk of suffering serious harm. Such a definition would allow those assessing performance to determine in individual cases whether the alleged deficiencies do, in practice, expose patients to risk.

The task of devising assessment procedures also raises difficult questions. First is the breadth of the assessment: should it cover the whole range of knowledge, skills, and attributes which might be needed by a doctor in a particular specialty or should it focus on those which have been the subject of a complaint? What methods of assessment are appropriate? And how can the assessments be tailored to doctors in different grades or with varying responsibilities?

Internal GMC working papers initially suggested that a comprehensive assessment would be attempted using a range of techniques, including a detailed proforma for the doctor to complete, multiple choice examinations, tests of a variety of skills by simulation, review of medical records, discussion of cases, structured interviews, and discussions with colleagues and managers. The appropriateness of some of these methods was questioned at a GMC internal conference in February on grounds of both relevance and reliability. Extensive biographical data, for example, would shed little light on the doctor's current standard of practice. And many doctors who are performing poorly might none the less be able to pass multiple choice examinations set at the very basic level envisaged by the council's assessment working groups.

These difficulties highlight the fact that assessments of performance cannot be conducted in a vacuum. Assessors will need to take account of the nature of the job being done by the doctor and the responsibilities that this entails. Only then can an appropriate assessment be devised. That assessment should focus on areas of alleged deficiency and should aim to determine what has been the doctor's level of performance in the recent past, rather than his or her ability to reach some standard under test conditions. Review of medical records and practice arrangements and discussion of specific cases should thus be the principal tools. Assessors should be given discretion to match the scope and methods of their inquiry to the circumstances of each case, within agreed guidelines, rather than attempting a cumbersome and questionable overall assessment. They should, of course, be free to broaden their inquiries if evidence suggests that this would be appropriate. Above all, assessors should keep constantly in mind the criterion against which they are measuring performance—namely, whether patients are at risk of suffering serious harm.

In this process the involvement of lay people will be essential, both to bring a patient's perspective to the assessments and to ensure public confidence in the procedures, but the specialised knowledge required to assess doctors' performance makes it inevitable that the process should essentially be one of peer review. This can succeed only with the support of the medical profession as a whole—including a willingness on the part of doctors to participate in the procedure as assessors. That support will depend on an understanding that the procedures will be concerned only with serious deficiencies and that the methods of assessment will be fair and reasonable.

STEPHEN BREARLEY
Consultant surgeon (and member of the GMC)

Whipps Cross Hospital,
London E11 1NR

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Primary biliary cirrhosis: epidemiology helping the clinician

Epidemiology improves our knowledge of disease

Recent epidemiological and clinical research is changing our thinking on primary biliary cirrhosis, a chronic cholestatic liver disease probably of autoimmune origin. Until the early 1970s primary biliary cirrhosis was perceived as a disease that was rare, presented with persistent jaundice, and usually progressed to death from liver failure.¹ Epidemiological and clinical research based on both registers of cases in defined populations and, more commonly, clinical case series has transformed that picture. The disease is much commoner than was believed,^{2,4} as many as half of all patients may have no symptoms of liver disease at diagnosis,⁵ and both the onset of complications and death can be delayed by treatment with ursodeoxycholic acid⁶ and, when necessary, liver transplantation. Good clinical and health care strategies will depend on further integration of epidemiological and clinical research to provide better information on the frequency and natural course of the disease and to generate hypotheses about causation.

For both epidemiological and clinical purposes a widely accepted and validated case definition is necessary. In primary biliary cirrhosis the triad of results from liver function tests

suggesting cholestatic liver disease, the presence of antimitochondrial antibody on indirect immunofluorescence, and compatible or diagnostic liver histology provide such criteria.⁷ A possible case may be regarded as having any two of these. Recent descriptive studies using these criteria have indicated an apparent dramatic increase in the prevalence of primary biliary cirrhosis in some regions but with considerable geographical variation. This variation is not only global but also extends to adjacent, apparently similar, countries such as Denmark, where prevalence is low, and Sweden, where it is high.^{2,8,9} North east England has seen an apparent tenfold increase in prevalence over the past 20 years.^{3,4}

It is still unclear to what extent these geographical and temporal variations, seen in a few areas where data are available, are due to true variations in prevalence or to variations in case registration, diagnostic activity, access to expertise in hepatology, or even local interest in the disease. These variations are important, both clinically, for health care planning, and for research. If there are true geographical variations in prevalence then different levels of awareness and investigation are needed according to local patterns. If, however, the geographical

variations are artefactual then clinicians in some areas are missing cases. A genuine rise in disease frequency requires planning now for the increased health resources needed in the future.

Most published studies have described case series and have not used consistent methods for case finding or screened a well defined population over a defined period. The use of an epidemiological framework¹⁰ to establish whether prevalence is truly increasing and to study geographical variation will require collaborative studies between clinicians and epidemiologists both locally and internationally, using registers based on internationally agreed protocols.

Evidence of true temporal and geographical variations in the prevalence of primary biliary cirrhosis will enable us to generate new hypotheses about its cause. Here analytical epidemiology can be combined with laboratory and clinical observations to help establish the cause. Many putative causes and risk factors for primary biliary cirrhosis have been suggested over the years. These have included an unknown factor associated with the water supply,¹¹ urinary tract infections caused by *Escherichia coli*, drugs (particularly phenothiazines), a history of obstetric complications or gynaecological interventions, disease onset after childbirth or anaesthetic,¹² and, most recently, an atypical mycobacterial infection (a response possibly associated with immune mimicry).¹³ None of these has been confirmed as having a direct or indirect causal relation.

Recent genetic studies have suggested that possession of one of the HLA class II antigens associated with the DRW8 locus may confer increased susceptibility to the disease, and it may be no coincidence that this relatively rare tissue type is more prevalent in populations thought to have the highest prevalence of the disease.¹⁴ At present a best guess as to cause is that a number of possible trigger factors may initiate progress of the disease in a susceptible individual. Furthermore, familial primary biliary cirrhosis (usually mother-daughter pairs) is described in 1-4% of cases,^{15 16} and other autoimmune diseases such as thyroid disease and rheumatoid arthritis are commonly found in patients with primary biliary cirrhosis. Surprisingly, there have been no well conducted case-control or cohort studies to examine these or other possible risk factors, and these hypotheses need testing. An understanding of the cause of primary biliary cirrhosis will enable clinicians to develop preventive programmes and rational treatment policies to evolve and answer patients' questions about the disease.

In conclusion, recent descriptive epidemiological studies have shown that primary biliary cirrhosis is by no means rare, certainly in northern Europe and particularly among middle aged women. Its rising prevalence, together with the fact that it may be present with few symptoms for months or years and that its cardinal symptom of lethargy is non-specific, should alert physicians and family doctors to its possible diagnosis in

any patient with a positive test result for antimitochondrial antibody, a highly specific serological marker for the disease. Our evolving understanding of the clinical course of the disease and an increasing range of treatment options mean that purchasers and providers of health care need to be aware of their changing obligations to these patients, including giving them access to the highly successful patient support group (the PBC Support Group). Continuing collaboration between epidemiologists and clinicians will provide important clues about the cause and pathogenesis of this enigmatic disease and develop our knowledge of its prevention and management.

The PBC Support Group may be contacted at the British Liver Trust, Central House, Central Avenue, Ransomes Europark, Ipswich IP3 9QG.

JANE V METCALF
Wellcome research fellow

Department of Medicine,
University of Newcastle upon Tyne,
Newcastle upon Tyne NE2 4HH

DENISE HOWEL
Senior lecturer in statistics

Department of Public Health and Epidemiology

OLIVER F W JAMES
Professor and head

Department of Medicine

RAJ BHOPAL
Professor and head

Department of Public Health and Epidemiology

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