

- 4 Firth-Cozens J. Emotional distress in junior house officers. *BMJ* 1987;295:533-5.
- 5 Allen I. *Any room at the top? A study of doctors and their careers*. London: Policy Studies Institute, 1988.
- 6 Dillner L. The road to disenchantment. *BMJ* 1992;305:1103.
- 7 British Medical Association. *Stress and the medical profession*. London: BMA, 1992.
- 8 Harden RM, Sowden S, Dunn WR. Some educational strategies in curriculum development: the SPICES model. *Med Educ* 1984;18:284-97.
- 9 Elton L, Partington P. *Teaching standards and excellence in higher education. Developing a culture for quality*. Sheffield: Committee of Vice Chancellors and Principals of the Universities of the United Kingdom, 1992.
- 10 Secretaries of State for Health, Wales, Northern Ireland, and Scotland. *Working for patients*. London: HMSO, 1989. (Cmd 555.)
- 11 Universities Funding Council Medical Committee. *First report on the effects of the NHS reforms on medical and dental education and research*. London: UFC, 1991.
- 12 Universities Funding Council Medical Committee. *Second report on the effects of the NHS reforms on medical and dental education and research*. London: UFC, 1992.
- 13 General Medical Council. *Recommendations as to the medical curriculum*. London: GMC, 1957.
- 14 General Medical Council. *Recommendations on basic medical education*. London: GMC Education Committee, 1980.
- 15 McManus C, Lockwood D. Medical education, training and research. In: Beck E, Lonsdale S, Newman S, Patterson D, eds. *In the best of health? The status and future of health care in the UK*. London: Chapman and Hall, 1992.
- 16 Stocking B. *Medical advances. The future shape of acute services*. London: King's Fund, 1992.

## London after Tomlinson

### Maintaining excellence: the preservation and development of specialised services

Liam J Donaldson

*This is the fourth article in our series looking at the issues highlighted by the Tomlinson report into London's health care and medical research and education*

**The advent of the Tomlinson inquiry draws attention to the need to strike a balance between market led and planned approaches to health care delivery. This is important not just for hospital rationalisation but also for the preservation and development of services which are provided in a smaller number of hospitals. Specialised services are often in the forefront of raising standards of care and introducing new developments and innovations. They are the only option for a small number of patients with serious illnesses. In the internal market for health care provision created by the 1990 NHS reforms more sophisticated and flexible mechanisms must be found to provide stability for specialised services while at the same time enabling the benefits of purchaser choice and provider competition to be realised.**

#### Introduction

The introduction of health care markets was one of the fundamental pillars of the white paper *Working for Patients*.<sup>1</sup> The proposals and subsequent legislation contained several key ingredients: the creation of an environment of competition among providers of services with the aim of reducing costs and raising quality; greater choice for, and more emphasis on, consumers of health services; greater accountability of professionals; and funding mechanisms which rewarded services sought after by patients and referring general practitioners.

In the early days of the NHS reforms purists ruled. The word "planning" was all but expunged from the lexicon of health service management and the former attempts of the NHS to plan in detail the nature and location of services were viewed as outmoded. There was complete faith that the forces released in the new market would both accomplish rationalisation of services not required to meet the needs of local populations and enable the emergence of new and improved services. It rapidly became apparent that the existence of entirely untrammelled markets would inevitably mean that some clinical departments and even some whole hospitals would quickly fail to retain a foothold in the new system of care. The spectre of bed closures and service rationalisation on a large scale would have been as controversial as it was under the old arrangements and would hardly have struck the public as a beneficial aspect of reforms that were working in their ultimate interests.

Together this general context and the scale of

#### Potential functions of a specialist centre

- Service delivery
- Research
- Innovation
- Postgraduate training
- Benchmark for good practice
- Advice and devolution of skill

potential problems in London led to the Tomlinson inquiry.<sup>2</sup> Its very establishment, however, raises questions about future mechanisms for change and development of services in the National Health Service. Aside from its specific recommendations for London, and its predictions about the appropriate balance between primary and hospital care, the importance of the Tomlinson report lies in whether the approach signals a return to planned solutions for difficult problems as distinct from purely market led change.

#### Planned and market led approaches

Striking an appropriate balance between planned and market led approaches is as relevant to considering how best to preserve and develop those specialised services which are concentrated in a small number of hospitals as it is to rationalisation. Concern has constantly resurfaced, particularly in professional circles, about the ability of the new market system to preserve highly specialised services or foster new ones. In neonatal care, for example, fears expressed by witnesses to the parliamentary select committee on health that the impact of the internal market will lead to a fragmentation of regional services and devolution of expertise into a multiplicity of small provider units led the committee to conclude: "We are not persuaded that the establishment of contracts for regional services for perinatal and neonatal intensive care can be left to market forces and audit."<sup>3</sup>

Under the new arrangements the configuration of specialist services will be determined ultimately by whether purchasers see a need for them and then through the resulting contracting mechanisms to secure and fund the care required. In theory there should be no difficulty. The purchasers of care (district health authorities and fundholding practices) are responsible for assessing and meeting the needs of their populations. Their decisions should enable people with clinical need who require services not available in a local hospital to receive them. However, the process through which need results in a decision to purchase is

**Northern Regional Health Authority, Newcastle upon Tyne NE6 4PY**

Liam J Donaldson, regional general manager and professor of applied epidemiology

*BMJ* 1992;305:1280-3

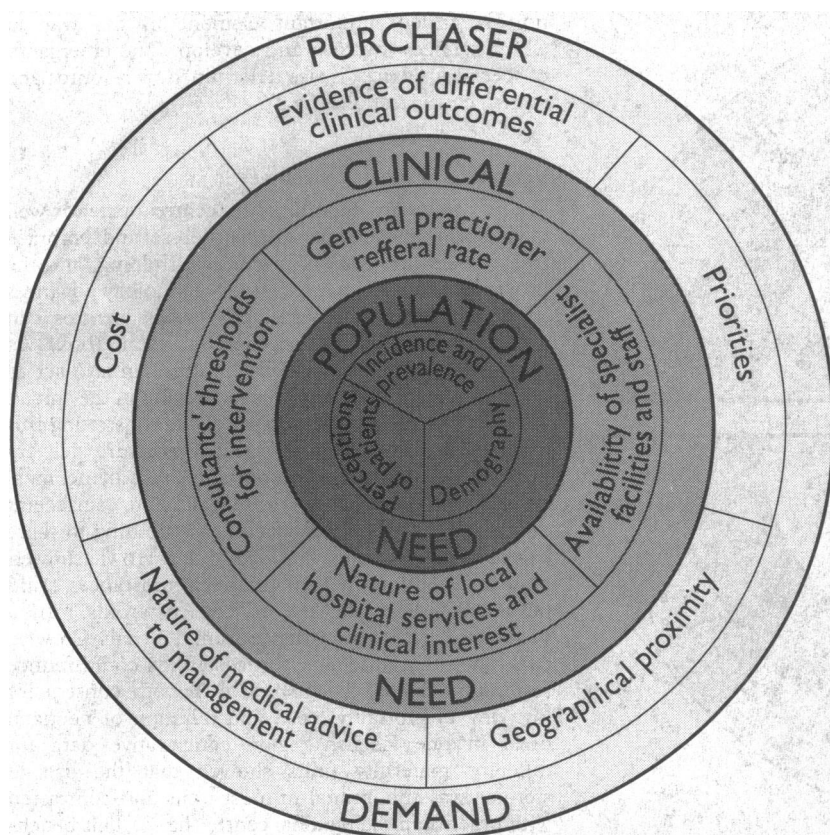


FIG 1—Main factors influencing demand for more specialised hospital services

TABLE I—Referral rates (per million population) and estimated tertiary referrals (%) for hepatobiliary services from health districts of a region

District	Rate	% Tertiary referrals
A*	2200	8
C	1000	18
L	790	23
B	670	27
I	450	40
F	310	58
J	230	78
N	160	100
G	150	100
D	150	100
K	120	100
H	50	100
E	30	100
M	10	100
Region	480	37

\*District in which service is located

TABLE II—Rates of admission (per million population) for coronary artery bypass grafting within populations of health districts in a region

District	Rate	Ratio to regional rate
A*	377	1.5
B	371	1.5
C	353	1.4
D	323	1.4
E	280	1.1
F	280	1.1
G	258	1.0
H	257	1.0
I	254	1.0
J	216	0.8
K	203	0.8
L	192	0.8
M	155	0.6
N	77	0.3
Region	256	1.0

\*District in which service is located

complex. It involves the extent of need at population level, how that is translated into clinical need, and what the purchaser then chooses to do about it. Each of these stages is subject to multiple influences (fig 1) which will vary from service to service and locality to locality.

If all specialised services were accessed through so called "tertiary" referrals (one consultant passing clinical responsibility to another) then clinically defined need would be the main influence shaping service configuration. This would, however, shift the balance away from the purchaser as the prime decision maker for the health care requirements of the local population, opening the door to a provider led determination of priorities, overemphasising complex services, and increasing costs in a way which the reforms were intended to prevent. On the other hand, many people within hospitals providing specialised services doubt the capacity of the new system to recognise bona fide need, believing the purchasing function to be too devolved, too fragmented, and too lacking in specialist medical advice. We must also remember that while in theory many specialist services are tertiary they can receive a substantial proportion of their work as referrals from general practitioners. This is seen, for example, in the analysis of the work of a specialist hepatobiliary service proposing to establish a liver transplantation service (table I).

A further important issue in considering the two approaches is how equity of access to health services will be affected. The Royal Commission on the National Health Service gave as one of the five overall objectives for the service providing "equality of entitlement to health care."<sup>4</sup> Over the years fulfilling this objective has proved problematical, especially in the provision of specialised services. A typical illustration is shown in table II. Residents of districts in a health region experience very different admission rates for coronary artery bypass grafting, a service provided in one hospital but serving the whole region. The highest admission rate was for residents of the district (A) where the regional centre was based and among the

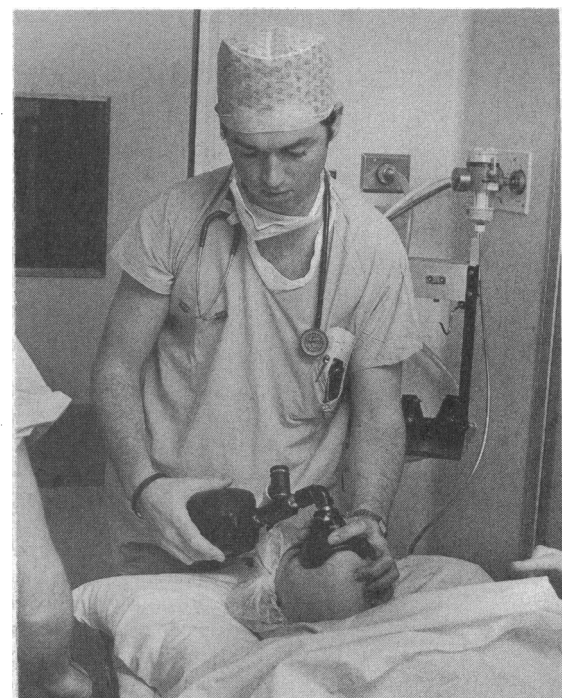
lowest for residents of a district (L) six miles away. Moreover, districts E, M, and N are part of the same conurbation but have very different admission rates. Interpreting such data is made difficult by the absence of a real understanding of what constitutes need—so it is not possible to say with certainty what level of admission would have been appropriate. Nevertheless, the known mortality pattern for coronary heart disease in the region concerned suggests that differing morbidity could not explain the variation in operation rates. Likely explanations include differences in clinical decision thresholds on the part of general practitioners and general physicians, the availability of local cardiology services, and distance from the regional centre.

If equity is to remain a fundamental value of the NHS, and inequalities like those shown are to be reduced, then this poses enormous conceptual and logistical challenges to the new purchasing function, the principal mechanism of change.

### Specialised services and how they develop

In introducing the concept of a district general hospital the 1962 hospital plan recognised the importance of linking it with centres providing regional specialist services,<sup>5</sup> an approach subsequently reinforced by the Bonham-Carter committee.<sup>6</sup> At this stage specialised services were generally defined in terms of clinical specialties—for example, neurosurgery, orthodontics, ophthalmology.

The modern concept of specialised services is much wider. Today such services differ according to their degree of geographical dispersal: some are provided in only one or two centres in England (heart transplantation), some in one or two centres in a health region (forensic psychiatry), and some in several centres in a health region (radiotherapy). They also differ in the way in which the specialist role is delivered. A specialised service can simply be a tertiary referral by a consultant to a colleague with particular skill. For example, a patient with severe rheumatoid arthritis might be referred by a rheumatologist to an orthopaedic surgeon who is an expert in hand surgery. Between-consultant referrals themselves vary greatly. Some consultants become particularly expert in carrying out one particular operation and some colleagues



Tertiary referral may simply be to a colleague with particular skills

JOHN TWINNING

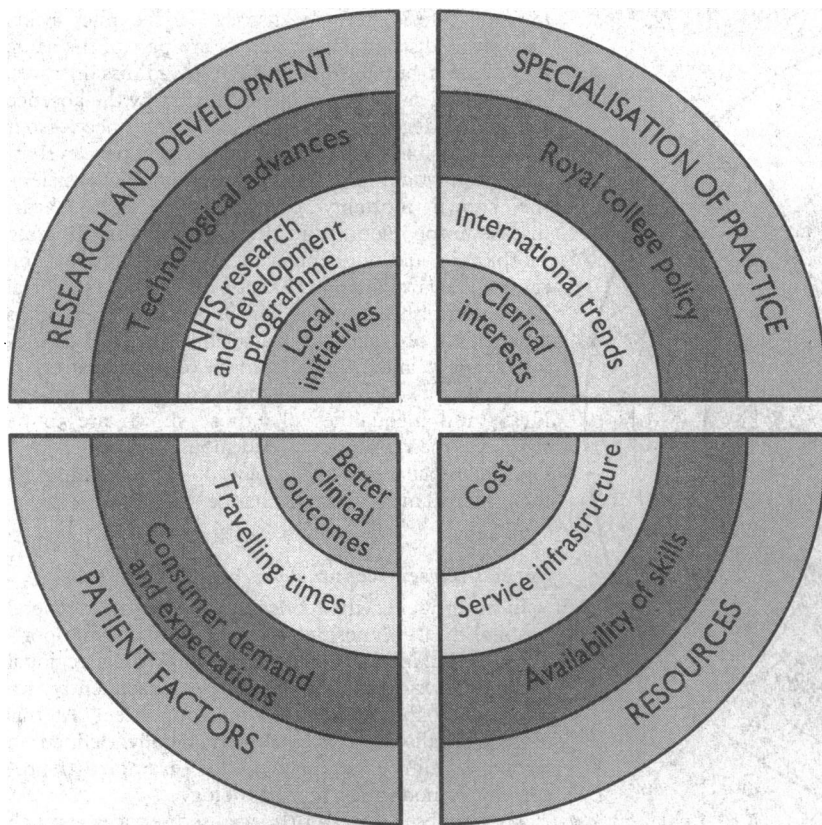


FIG 2—Emergence of new specialised services: underlying forces

may pass all such cases over to him or her. This form of specialised service within a general case load usually arises because of better clinical outcome in the hands of a particularly gifted individual or skilled team. Other organisational models for specialist services still involve whole specialties (for example, paediatric neurology), though the number of such specialties has increased; single diagnostic procedures (for example, drug assays); and the treatment of a particular condition (for example, breast cancer).

The emergence of new specialised services is prompted by a potentially wide range of influences (fig 2). Some concern greater specialisation in clinical practice and the extent to which better outcome is achieved by specialist centres. In interpreting studies which appear to show better results for patients treated in specialist centres it is important to remember that volume could affect outcome for two entirely separate reasons: "practice makes perfect" or "selective referral."<sup>7</sup> Important decisions about the organisation and development of services will always have to be taken because of changes in the degree of specialisation in clinical practice. This is illustrated by the position of vascular surgery, which is close to the point of emerging as an independent subspecialty of general surgery. If no overview is taken of the organisation of such services within a region there is a possibility that purchasers may decide to build up potentially less effective local services to avoid the cost of referral to specialist centres.

New discoveries, advances in technology, and innovation are further influences in the emergence of new services. The NHS is developing a strong centrally led research and development function which will be very important in shaping the development of specialised services.<sup>8</sup> It is also addressing the issue of systematically evaluating new technologies in a way which has never previously been done.<sup>9</sup> This will go some way to assuaging the fears of those who see clinician led, unplanned introduction of new technologies ("creeping growth") as one of the most potent sources of the health service's financial difficulties. Nevertheless, it is important to remember that clinical innova-

tion is a very important element in the way in which services improve and develop. The emergence of specialist services is also determined by resource and patient considerations (fig 2).

### Functions of specialist centres

The functions of specialist centres extend well beyond the delivery of services per se and embrace research and innovation in practice, training, and the provision of advice. Moreover, as technology advances some services which begin in specialist centres can eventually be devolved to general hospitals (box). As centres of excellence specialist centres can also act as benchmarks for other services in a region in the spread of best practice into routine use. Data illustrating this function are shown in figure 3. Premature delivery remains the single most important cause of neonatal death, with respiratory distress due to surfactant deficiency being the main factor contributing to these deaths.<sup>10</sup> Twenty years ago Liggins and Howie showed that surfactant deficiency in these circumstances could be prevented by the use of corticosteroids.<sup>11</sup> In a systematic review of evidence from 12 trials Crowley concluded that antenatal corticosteroids could reduce respiratory distress by 40% to 60% with consequent lowering of mortality, cost, and duration of neonatal care. In one English region comparative data for different maternity units showed that the use of steroids was suboptimal in most units and contrasted with practice in the regional centre (fig 3). This benchmarking exercise allowed all units to review their practice and change their clinical policies. Change has implications not just for quality of care but for its cost since corticosteroid administration is cheap compared with the alternative of neonatal intensive care.

### Contracting mechanisms

Ultimately the preservation and development of specialised services will depend on how well the contracting system can address the particular nature of "high cost, low volume" services. Present methods vary. Some specialised services are covered by routine

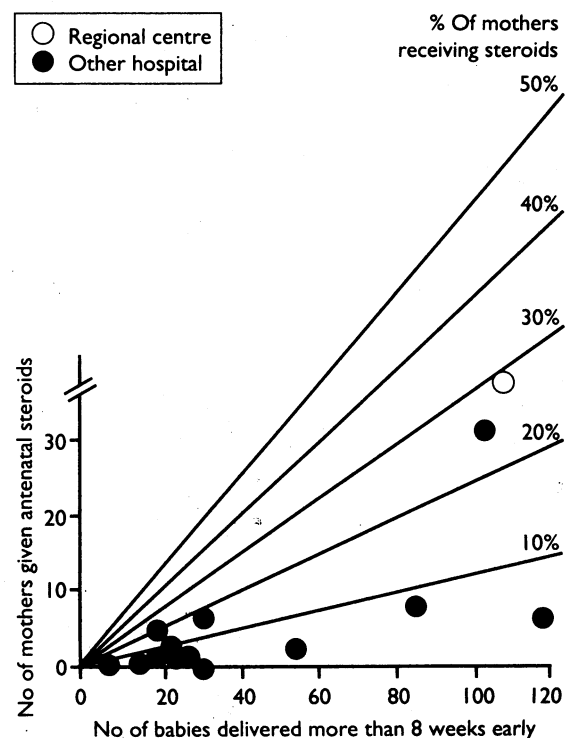


FIG 3—Use of steroids in differing maternity units in an English health region. Source: Khanna and Richmond<sup>10</sup>

block or cost and volume contracts, but small numbers and a consequent high degree of fluctuation in case numbers make these difficult to set and monitor for individual purchasers. Some are covered by cost per case contracts or are left to be picked up as extra-contractual referrals, but this approach offers no assured revenue (and therefore no longer term stability) to hospitals providing specialised services.

Current thinking appears divided between a mechanism based on contracts continuing to be placed by district health authorities for tertiary referrals and a system of automatic retrospective reimbursement (with attendant loss of financial control for purchasers and perverse incentives for the referring hospital). Neither of these mechanisms is likely to be wholly satisfactory, and a more sophisticated approach is needed. Some possible examples are shown in table III, including the "insurance-premium" purchaser contract, and the provider subcontract for tertiary referrals within a secondary service commissioned as a single whole by purchasers.

## Conclusion

The new internal market for public health care provision is now in its second full year of operation. It has brought undoubted benefits: it has made explicit considerations of quality and cost; it is challenging assumptions about the balance between primary care and hospitals and it is enabling decisions about resource allocation to be governed by need, not purely demand. Nevertheless, there are some issues which the internal market and the mechanism of contracting cannot resolve unaided. It is important that a strategic overview is taken and a direction for change set out.

Tomlinson has shown how this can be successfully achieved for major service rationalisation without compromising the benefits of the internal market in the longer term.<sup>2</sup> Specialised services are one of the most precious commodities of the health service. They often lead the way in quality and provide ideas and innovation for the future. For a small number of patients they may be their only source of help. New mechanisms must be found to ensure that they

TABLE III—Range of potential contracting models for specialist services and its use in one health region

Contract model and its features	Examples of current or potential application
<p><i>Supraregional finance</i></p> <ul style="list-style-type: none"> <li>● High cost/low volume services meeting rigorous criteria</li> <li>● National/cross regional catchment area</li> <li>● Services centralised in a few providers</li> <li>● Subject to overall workload, DHAs have guaranteed access</li> <li>● "Insurance premiums" paid via top slicing of RHA allocations</li> </ul>	<ul style="list-style-type: none"> <li>● Heart and lung transplants</li> <li>● Liver transplants</li> <li>● Neonatal and infant cardiac surgery</li> </ul>
<p><i>Cross regional insurance contracts</i></p> <ul style="list-style-type: none"> <li>● High cost/low volume services centred on a few hospitals with cross regional catchment area</li> <li>● For services which do not meet supraregional criteria or awaiting designation</li> <li>● Volatility of referrals such that "risk pool" needs to cover several regions</li> <li>● Can be simple block contracts collectively financed by DHAs in several regions or more sophisticated insurance arrangements between the DHAs and the hospital</li> </ul>	<ul style="list-style-type: none"> <li>● Forensic psychiatry (especially court referrals)</li> <li>● Head injury/severe brain damage</li> <li>● Neonatal extracorporeal membrane oxygenation</li> <li>● Haemophilia (the very high cost cases)</li> <li>● Prolonged high cost intensive care and other "big" ECRs</li> </ul>
<p><i>Intraregional insurance</i></p> <ul style="list-style-type: none"> <li>● High cost/low volume services centred on few hospitals in region with a supradistrict catchment</li> <li>● Volatility of referrals such that risk is manageable within the region</li> <li>● Simple "as required cost sharing" agreement between several DHAs or can be more sophisticated "insurance premiums" in advance by DHAs to the provider</li> <li>● Arrangements need to be explicit on how much of any costs/excess costs are borne by (a) the hospital's contract income, (b) the DHA of residence, (c) other DHAs, (d) RHA brokerage</li> </ul>	<p>Being developed in the Northern region—though at present tends to be more of an ad hoc response than application of established insurance procedures.</p> <p>Has proved valuable in:</p> <ul style="list-style-type: none"> <li>● 2 separate high cost haemophilia cases at a local teaching hospital</li> <li>● an out of region referral by the courts for forensic psychiatric assessment/treatment</li> </ul>
<p><i>RHA purchasing</i></p> <ul style="list-style-type: none"> <li>● High cost/specialist services centred on a few hospitals in region with a regional catchment</li> <li>● Subject to overall workload all DHAs in region have guaranteed access</li> <li>● DHAs "insurance premiums" paid by top slicing of their allocations by RHA</li> </ul>	<p>Not widely used because (a) not part of RHA "core" role and (b) disenfranchises DHAs from responsibility to purchase the total health needs of their residents</p> <p>May remain in some regions as transitional arrangement/legacy of former "regional specialities"</p>
<p><i>Regionwide block contracts</i></p> <ul style="list-style-type: none"> <li>● High cost/specialist services centred on a few hospitals in region with a regional catchment</li> <li>● Subject to overall workload, all DHAs in region have guaranteed access "free" at point of use</li> <li>● DHAs' "insurance premiums" paid by contributions made by each DHA (for example, on basis of weighted population) to a lead purchaser who negotiates/runs contract on their behalf</li> <li>● Hospitals have guaranteed income for much of their costs and economy of effort in negotiations</li> <li>● Hospitals may also have separate contracts/ECRs from non-participating DHAs for the service</li> </ul>	<ul style="list-style-type: none"> <li>● Spinal injuries (for hospitals both within and outside the region)</li> <li>● Regional rehabilitation service</li> <li>● Communication aid centre</li> <li>● Regional genetics service</li> <li>● Secure/semisecure facilities for learning disabilities and mental illness</li> <li>● Supraregional/regional assay service (for hospital both within and outside the region)</li> </ul>
<p><i>Coordinated contracts</i></p> <ul style="list-style-type: none"> <li>● High cost/specialist services centred on a few hospitals in region with a regional catchment</li> <li>● Separate contracts negotiated by individual DHAs/consortia to ensure (a) consistency of service, (b) economy of effort in negotiation, (c) common approach, (d) avoidance of differential access/standards or all contract negotiations coordinated by a steering group representing all DHAs and the hospital</li> </ul>	<ul style="list-style-type: none"> <li>● Specialist radiotherapy services</li> <li>● Neurosciences</li> </ul>
<p><i>Consortium contracts</i></p> <ul style="list-style-type: none"> <li>● High cost/specialist services centred on a few hospitals in region with a subregional catchment</li> <li>● DHA groups collectively negotiate contracts for (a) economies of scale/effort, (b) bulk purchasing, (c) develop purchaser expertise, (d) "insurance" against volatility of each DHA's referrals</li> </ul>	<ul style="list-style-type: none"> <li>● Specialist psychiatry</li> <li>● Cardiothoracic surgery</li> </ul>
<p><i>Umbrella contracts</i></p> <ul style="list-style-type: none"> <li>● Useful for DHAs/fundholders for small volumes of referrals in several specialties from a single hospital</li> <li>● Referrals (often of similar cost) in several specialties grouped together under a single "umbrella"</li> <li>● Flexibility between the specialties involved for purchaser, stable income for hospital</li> </ul>	<p>Tends to be used by DHAs buying cases from remote hospital and fundholders purchasing a package of small individual volumes</p>
<p><i>Individual contracts</i></p> <ul style="list-style-type: none"> <li>● The "lowest" form of contract between a single DHA and single provider covering one specialty or subspecialty</li> </ul>	<p>Widespread use</p>
<p><i>Provider subcontracts</i></p> <ul style="list-style-type: none"> <li>● Purchaser contract for a "whole service," including costs of subsequent tertiary referral for a proportion</li> <li>● Removes incentive for provider to refer on to escape costs</li> <li>● Tertiary provider security and fixed costs still imply need for consortia/insurance premium approach by secondary providers if low volume service</li> </ul>	<p>Not yet common, but examples include:</p> <ul style="list-style-type: none"> <li>● Rehabilitation services</li> <li>● Patient transport services</li> </ul>

RHA=regional health authority; DHA=district health authority; ECR=extracontractual referral.

are developed and maintained while preserving the benefits of the new system of health care.

These challenges confront all regions in the country but are especially important in London. As hospitals in the capital are reconfigured, as services currently located in specialist hospitals which are perceived as too dispersed are relocated, and as some skilled staff take the opportunity to bow out of the service, it is essential that the present and future place of specialised services are secured within the network of care.

I am grateful to my colleagues Bill Kirkup and Tim Watkinson for the thinking which resulted in the contract matrix shown in table III.

- 1 Secretaries of State for Health, Wales, Northern Ireland and Scotland. *Working for patients*. London: HMSO, 1989. (Cmnd 555.)
- 2 *Report of the inquiry into London's health service, medical education and research*. London: HMSO, 1992.

- 3 House of Commons Health Committee. *Second report. Maternity services*. London: HMSO, 1992. (HC 29-1.)
- 4 Royal Commission on the National Health Service. *Report*. London: HMSO, 1979. (Cmnd 7615.)
- 5 *A hospital plan for England and Wales*. London: HMSO, 1962. (Cmnd 1604.)
- 6 Central Health Services Council. *The functions of the district general hospital*. London: HMSO, 1969.
- 7 Roberts H. *Outcome and performance in health care: a guide to developments in outcome and performance assessment in the NHS*. London: Public Finance Foundation, 1990.
- 8 Department of Health Research and Development Division. *Research for Health: a research and development strategy for the NHS*. London: Department of Health, 1991.
- 9 Advisory Group on Health Technology Assessment. *Assessing the effects of health technologies: principles, practice, proposals*. London: Department of Health, 1992.
- 10 Khanna R, Richmond S. Antenatal treatment to enhance maturity. In: *Report on perinatal and late neonatal death in the Northern region 1991*. Newcastle upon Tyne: Northern Regional Health Authority, 1992.
- 11 Liggins GC, Howie RN. A controlled trial of antepartum glucocorticoid treatment for prevention of the respiratory distress syndrome in premature infants. *Pediatrics* 1972;50:515-25.
- 12 Crowley P. Promoting pulmonary maturity. In: Chalmers I, Enkin M, Keirse MJNC, eds. *Effective care in pregnancy and childbirth*. Oxford: Oxford University Press, 1989:125-30.

## Using a mock trial to make a difficult clinical decision

Richard Smith

**The motion: Bone marrow transplantation should forthwith be offered to all children in the UK with symptomatic sickle cell anaemia who have HLA matched siblings.**

Many clinical decisions have to be taken with inadequate scientific information. Reaching a consensus among experts has been tried as one response to this problem. Another, described here, is to use legal process to dissect a difficult question. In this case a mock trial—using barristers, expert witnesses, and a jury—was conducted on whether bone marrow transplantation should be offered to all children with symptomatic sickle cell disease. Transplantation seems to offer about a 90% cure rate for a condition that may kill 15% of children before they reach 20. But transplantation carries a 10% risk of death or severe disability, and doctors cannot predict which children will suffer severely from their sickle cell disease and which will suffer little or nothing. The jury eventually reached a majority decision that transplantation should not be offered now to all symptomatic children.

Haematologists cannot agree whether children with symptomatic sickle cell disease should be offered bone marrow transplantation. Preliminary evidence suggests that it may be the only “cure” for this potentially fatal condition<sup>1,2</sup>: the transplanted marrow replaces the diseased marrow and the child thereafter produces normal haemoglobin. Some haematologists want to offer bone marrow transplantation to children with symptomatic disease—despite the fact that bone marrow transplantation itself may kill or disable. Others think it premature to offer the treatment so broadly and that much more evaluation needs to be done first.

Sally Davies, a haematologist from the Central Middlesex Hospital, faces the dilemma of whether to offer such treatment and recently attended a National Institutes of Health workshop on the subject.<sup>3</sup> Consensus could not be reached. She wanted help with her dilemma and thought that she might look to legal methods. After all, the courts are daily having to sift through evidence and make judgments on what look like impossibly difficult problems; and Dr Davies had been impressed by how cross examination by barristers when she had appeared as an expert witness had helped her clarify her own thinking on difficult issues. Another advantage of trials is that they must be conducted in ordinary language, allowing patients and others to be brought into the debate.

Dr Davies set up a mock trial on the use of bone

marrow transplantation in the Middle Temple. She managed to persuade three Queen's Counsels and four eminent haematologists, two of them from the United States, to participate in the trial. In addition, she assembled a jury of non-experts: a nurse with sickle cell disease, a mother whose child had died of the disease, a black community activist, a black barrister, a black junior doctor, a regional medical officer, and the editor of a general medical journal (me).

We had six hours to argue it out in the wood panelled hall under five gigantic pictures of kings of England. The motion was that “Bone marrow transplantation should forthwith be offered to all children in the UK with symptomatic sickle cell anaemia who have HLA matched siblings.”

### Case for the motion

Jean Richie QC opened her case by telling the jury and audience that some 5000 people in Britain have sickle cell disease; about 60-70% of these have the double recessive form of the disease to which the motion refers. In any given case the course is “totally unpredictable.” Patients may be unaffected by the disease, or they may die of it. Cross sectional, hospital based data from the United States show that 15% of patients with double recessive disease die before they are 20 and 50% before 40. Patients may die of infection, stroke, or organ failure, and they may suffer a wide range of problems, including vaso-occlusive crises, brain damage, impotence, and blindness.

Routine treatment includes prophylactic antibiotics and blood transfusion, but attempts to develop anti-sickling drugs have failed. One treatment that may prove useful is hydroxyurea, which encourages the production of fetal haemoglobin. Although this treatment is undergoing a controlled trial, the results are not yet available. Hydroxyurea is not recommended in children because of fear of long term side effects. Gene therapy may eventually be useful, but nothing is yet available. Prenatal diagnosis and termination of pregnancy is offered for double recessive disease, and uptake is about 50% in both Britain and the United States.

Bone marrow transplantation from an HLA matched donor is the only current “cure” for the condition. It has been used in some 40 patients—mostly Africans