

and kilopascals so that users become familiar with kilopascals. Finally, the medical and nursing professions, the clinical market for blood pressure measuring devices, must ensure that manufacturers provide us with accurate devices designed to our specifications, rather than accepting, as we have in the past, devices in which these considerations are secondary to the commercial success of the product.

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EOB is a member of the board of AccuSphyg LLC, New York, a company developing an automated device. He has also received funding over the past decade from several blood pressure device manufacturers to perform validation studies on automated devices, the results of which have been published in peer reviewed journals.

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Heart and heart-lung transplantation in Down's syndrome

The lack of supportive evidence means each case must be carefully assessed

Congenital heart disease is common in Down's syndrome, occurring in about 40% of individuals.¹ Twenty years ago cardiac surgery was often not attempted in children with Down's syndrome because of operative mortality of up to 60% and a short life expectancy.² With improvements in paediatric cardiac surgery and changes in attitude towards children with Down's syndrome such children now undergo corrective cardiac surgery. Some will inevitably develop complications and may benefit from heart transplant. There is also a large group of young adults with Down's syndrome who did not have heart surgery when young and who have uncorrected heart lesions that are now inoperable because of irreversible pulmonary vascular disease. They too are potential candidates for heart-lung transplantation. There is no published literature on heart or heart-lung transplantation in Down's syndrome, which makes it hard to predict the outcome in these patients.

Heart transplantation is now a widely accepted treatment, and medium term survival has steadily improved.³ The results of heart-lung transplantation are not as good but have also improved. Long term outcome of both is uncertain, with rejection and the side effects of immunosuppressive drugs (malignancy and infection) the major complications.

During a 14 year programme with over 800 transplants we have received only one referral for a patient with Down's syndrome. A questionnaire sent to other UK transplant centres revealed only two other referrals. None of these patients underwent transplantation (for reasons other than Down's syndrome). However, the paucity of referrals is surprising given the high prevalence of Down's syndrome and associated cardiac problems. A similar situation has been noted in

paediatric oncology, with a lower than expected number of referrals for bone marrow transplant.⁴

Although few people are consciously prejudiced, parents, referring physicians, and transplant centres may all worry that that a transplant will be "too much" for someone with Down's syndrome or that the patient will be difficult to manage. Coexisting medical problems are common and may be contraindications to transplantation. There is also concern over infective and malignant complications. Although no published work addresses the risks of heart transplant in Down's syndrome, some information can be drawn from literature about the immune system, bone marrow, and renal transplants in this population.

Well documented immunological abnormalities in Down's syndrome result in a high incidence of infection, autoimmune disease, and malignancy. Impaired chemotaxis, antibody production, phagocytosis, and bacteriocidal activity; reduced numbers of circulating lymphocytes; and an abnormal thymic structure are all recognised, although controversy exists on the precise immune defects.⁵ Immune abnormalities lead to an excess of all types of infection, but particularly to pneumonia (because of physical differences, including smaller airways, enlarged tonsils and adenoids, and lax muscle).¹

Leukaemia is 10-30 times more common in Down's syndrome. Reports on bone marrow transplantation describe increased infective complications, higher early mortality after transplantation, and more chemotherapeutic toxicity.⁶ Both the increased incidence of haematological malignancy and the increased sensitivity to chemotherapeutic agents may be due to a decreased ability to repair genetic damage, which has been shown in vitro.⁷ This has implications for the risk

of post-transplant malignancy secondary to long term immunosuppression. A report describing post-transplant lymphoproliferative disease in a normal recipient of bone marrow from a donor with Down's syndrome supports this.⁸ Post-transplant lymphoproliferative disease is usually a rare complication of bone marrow or renal transplantation. Its incidence is 5-10% after heart transplant, and this is likely to be much higher in recipients with Down's syndrome.

Apart from medical concerns about complications, there are broader ethical issues. All nine consultants who responded to our questionnaire said that patients' ability to understand the transplant process would influence the decision to accept them. Although there is an understandable reluctance to submit a mentally handicapped person to a process they cannot fully understand, many young children are transplanted after a decision is reached on their behalf with their family. Case reports of renal transplants in Down's syndrome^{9 10} and assessment of children undergoing bone marrow transplants¹¹ are encouraging and report no problems with compliance in their selected patients.

Although prejudice against transplant recipients on racial grounds has caused widespread condemnation, both the public and transplant specialists may be uncomfortable about allocating limited donor organs to patients with Down's syndrome, especially if they are considered "high risk" transplants. We also may be perceived as forcing patients to have transplants when they do not fully understand them. On the other hand, concern has recently been voiced that people with Down's syndrome may receive suboptimal medical care.¹² We need to ensure that such patients are treated fairly by the transplant community. We believe that each case should be considered on its merits, including an assessment of social support. The family should be made aware of the current lack of experience of transplantation in Down's syndrome and of our anxiety

about higher complication rates. If transplantation is still acceptable despite the unquantifiable risk, less aggressive immunosuppressive regimens should be considered, given underlying immune problems. The current paucity of referrals is unlikely to continue, and units should be prepared for the complex medical and ethical issues they will raise.

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Specialist registrar training

Some good news at last

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The headline news from the NHS has made grim reading this year: funding crises, shortages of beds, and crimes. The public is increasingly aware of our unenviable record of morbidity and mortality from cardiovascular diseases and cancer, and constructive criticism has been replaced by the destructive soundbite. The perception is of an NHS that has gone downhill since the halcyon days of the 1950s—overwhelmed by bureaucracy and initiatives that impede advances in clinical practice.¹

Yet there is another tale to tell. Over the past decade a quiet revolution has occurred in medical education. After the publication of the General Medical Council's recommendations in *Tomorrow's Doctors* in 1993² all UK medical schools have revised their undergraduate curriculums. Alongside a strong science base,³ *Tomorrow's Doctors* emphasised the importance of communication skills, learning through curiosity, understanding public health medicine, and adapting to changing patterns of health care. The burden of factual

information really was reduced, and a core curriculum defined. The implementation of these recommendations has not only influenced the medical students but also changed the way in which their seniors undertake both their teaching and their clinical practice.⁴

The culture of British medicine was already changing when the GMC issued *Good Medical Practice* in 1995, outlining the duties and responsibilities of a doctor.⁵ By emphasising the positive attributes of a doctor, that document has proved influential in defining the standards, including teaching and training, against which a doctor's professional performance can be assessed. The GMC then published recommendations for the preregistration house officer year in 1997⁶ and, with help from the departments of health, ensured their implementation. These developments in the UK were compatible with changes to medical education introduced in other countries.⁷

If these developments were broadly acceptable, the Calman reforms of specialist training proved more

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