- 1 Anderson W. A case of "Angeo-keratoma." *Br J Dermatol* 1898;10:113-7.
- 2 Fabry J. Ein Beitrag zur Kenntniss der Purpura haemorrhagicanodularis (Purpura papulosa haemorrhagica hebrae). *Archiv für Dermatologie und Syphilistherapie* 1898;43:187-200.
- 3 Kint JA. Fabry's disease: alpha-galactosidase deficiency. *Science* 1970;167:1268-9.
- 4 Desnick RJ, Sweely CC. Fabry's disease: alpha-galactosidase A deficiency. In: Stanbury JB, Wyngaarden JB, Fredrickson DS, Goldstein JL, Brown MS, eds. *The metabolic basis of inheritable disease*. New York: McGraw-Hill, 1983:906-44.
- 5 Kleiser WJ, Husaarts-Odijk LM, Sachs ES, Jahoda MGJ, Niermeiser MF. Prenatal diagnosis of Fabry's disease by direct analysis of chorionic villi. *Prenat Diagn* 1987;7:283-7.
- 6 Shows TB, Brown JA, Haley LL, Goggin AP, Eddy RL, Byers MG. Assignment of α -Galactosidase (α Gal) to the q22→qter region of the X chromosome in man. *Cytogenet Cell Genet* 1978;22:541-4.
- 7 Miller OJ, Siniscalco M. Report of the committee on the genetic constitution of the X and Y chromosomes. *Cytogenet Cell Genet* 1982;32:179-90.
- 8 Fox MF, Du Toit DL, Warnich L, Retief AE. Regional localisation of α -Galactosidase (GLA) to Xpter→q22, hexosaminidase B (HEXB) to 5q13→qter and arylsulphatase B (ARSB) to 5pter→q13. *Cytogenet Cell Genet* 1984;38:45-9.
- 9 Morgan SH, Cheshire JK, Wilson TM, MacDermot K, Crawford M d'A. Anderson Fabry disease—family linkage studies using two polymorphic X-linked DNA probes. *Paediatric Nephrology* 1987;1:536-9.
- 10 MacDermot KD, Morgan SH, Cheshire JK, Wilson TM. Anderson-Fabry disease, a close linkage with highly polymorphic DNA markers DXS17, DXS87 and DXS88. *Hum Genet* 1987;77:263-6.
- 11 Bishop DF, Desnick RJ. Maladie de Fabry. In: Funck-Brenato JK, Bach JF, Grunfeld JP, eds. *Actualités néphrologiques*. Paris: Flammarion Medicine-Sciences, 1988:99-108.
- 12 Wallace HJ. Anderson-Fabry disease. *Br J Dermatol* 1973;88:1-23.
- 13 Lockman LA, Hunninghake DB, Krivit W, Desnick RJ. Relief of pain of Fabry's disease by diphenylhydantoin. *Neurology* 1973;23:871-5.
- 14 Lenoir G, Rivron M, Gubler MC, Dufer JL, Tome FSM, Guivarch M. La maladie de Fabry. Traitement du syndrome acrodyneiforme par la carbamazépine. *Arch Fr Pédiatr* 1977;34:704-16.
- 15 Igarashi T, Sakuraba H, Suzuki Y. Activation of platelet function in Fabry's disease. *Am J Hematol* 1986;22:63-7.
- 16 Sakuraba H, Igarashi T, Shibata T, Suzuki Y. Effect of vitamin E and ticlopidine on platelet aggregation in Fabry's disease. *Clin Genet* 1987;31:349-54.
- 17 Goldman ME, Cantor R, Schwartz MF, Baker M, Desnick RJ. Echocardiographic abnormalities and disease severity in Fabry's disease. *J Am Coll Cardiol* 1986;7:1157-61.
- 18 Sakuraba H, Yanagawa Y, Igarashi T, et al. Cardiovascular abnormalities in Fabry's disease. A high incidence of mitral valve prolapse in hemizygotes and heterozygotes. *Clin Genet* 1986;62:285-7.
- 19 Maizel SE, Simmons RL, Kjellstrand CK, Fryds DS. Ten year experience in renal transplantation for Fabry's disease. *Transplant Proc* 1981;13:57-9.
- 20 Donati D, Navario R, Gastaldi L. Natural history and treatment of uraemia secondary to Fabry's disease: an European experience. *Nephron* 1987;46:353-9.
- 21 Popli S, Molnar ZV, Leehey DJ, et al. Involvement of renal allograft by Fabry's disease. *Am J Nephrol* 1987;7:316-8.
- 22 Brady RO, Tallman JF, Johnson WG, et al. Replacement therapy for inherited enzyme deficiency: use of purified ceramide trihexosidase in Fabry's disease. *N Engl J Med* 1973;289:9-14.
- 23 Desnick RJ, Dean KJ, Grabonski GA, Bishop DF, Sweely CC. Enzyme therapy. XII. Enzyme therapy in Fabry's disease. Differential enzyme and substrate clearance kinetics of plasma and splenic α -galactosidase isoenzymes. *Proc Natl Acad Sci USA* 1979;76:5326-30.
- 24 Touraine JL, Malik MC, Maire I, et al. Fetal liver transplantation in congenital enzyme deficiencies in man. *Exp Hematol* 1982;10(suppl 2):46.
- 25 Touraine JL, Malik MC, Traeger J, Perrot H, Marie I. Attempt at enzyme replacement by fetal liver transplantation in Fabry's Disease. *Lancet* 1979;i:1094-5.
- 26 Bishop DF, Calhoun DH, Bernstein HS, Hantzopoulos P, Quinn M, Desnick RJ. Human α -Gal A: nucleotide sequence of a cDNA clone encoding the mature enzyme. *Proc Natl Acad Sci USA* 1986;83:4859-63.
- 27 Bishop DF, Kornreich RA, Desnick RJ. Structure of the human α -galactosidase A gene: 5' control elements, intron/exon splice junction sequences, lack of propeptide sequence, and alternative 3' termination. *Am J Hum Genet* 1987;41(suppl):A208.
- 28 Susi ST, Martin BM, Kaslow DC, et al. Signal sequence and DNA mediated expression of human lysosomal α -galactosidase A. *Eur J Biochem* 1987;165:275-80.
- 29 Hantzopoulos PA, Calhoun DH. Expression of human α -galactosidase A in Escherichia coli K-12. *Gene* 1987;29:159-69.

HIV in prisons

Action, research, and condoms needed

Last month the Prison Reform Trust estimated that between 350 and 500 of the almost 50 000 prisoners in England and Wales are infected with the human immunodeficiency virus (HIV).¹ This high prevalence arises because drug abuse and probably homosexual sex are common among prisoners. The trust thinks that the prison authorities are failing in their response to HIV infection in prisons and wants, among other measures, the provision of condoms to male prisoners, an end to segregation of prisoners known to be infected with HIV, improved counselling and treatment for infected prisoners, a reassessment of the treatment of drug abusers in prisons, and extra training for prison medical officers.

As misusing intravenous drugs often leads directly or indirectly to imprisonment intravenous drug misusers constitute a high proportion of prison admissions. In Scotland in 1986, for example, the average daily prison population was 5588 and there were 521 admissions for drug offences related to drugs. In most penal institutions in the United States and Europe where studies have been undertaken infection with HIV is highest among intravenous drug misusers, and its prevalence is directly related to that in the local population of drug misusers. Thus a survey of inmates of prisons in New York State showed that 77 out of 494 men had antibodies to HIV (B I Truman *et al*, fourth international conference on AIDS, Stockholm, 1988).² Such data are not available for British prisons. Neither do we know how often needles are found in prisons.

The other risk of transmission of HIV within prisons is unsafe homosexual activity. Although it is generally believed that homosexual activity is common among prisoners, there are few data to substantiate this belief. Generally the prison population is young with pressing sexual urges that may be relieved only by self masturbation or homosexually, and many men whose sexual orientation is primarily heterosexual will take part in homosexual activity when they have no access to women. If prisoners participate in activities likely to put them at risk of infection with HIV they may after release act as a source of infection to the community. The types and

frequency of sexual activity participated in by men in prison have not been adequately investigated, and such information would clearly be important in determining policies.

Anecdotal evidence for genitourinary physicians who have treated rectal gonorrhoea in prisoners suggests, however, that anal intercourse does occur. In 1987 the World Health Organisation's special programme in AIDS stated that prison authorities must ensure the safety of prisoners and staff and minimise the further spread of HIV infection within prisons.³

In Britain much has been done to educate prison staff on minimising the further spread of HIV while undertaking their day to day duties. Minimising the further spread of HIV among the prisoners is more complex. In some countries such as Portugal all prisoners screened for antibodies to HIV and those who have the antibodies are isolated.⁴ This policy is not entirely safe because some infected people are seronegative. Within Europe there is considerable variation in how prisoners who are seropositive are managed: in some countries they are not subject to restrictions, but in others, including Britain, some separation is possible.⁴ Although routine serological testing for HIV infection is not undertaken in Britain, the segregation of prisoners who have engaged in high risk activities might reduce appreciably the risk of spreading the virus to others. Individual prisoners may, however, have acquired the infection in the past and stopped the risky activity, which makes them hard to identify.

The practicability of issuing sterile injecting equipment to prisoners has not been assessed adequately, and this matter clearly requires much attention. Education on the risks of injecting drugs should, however, be possible.

As our prisons are overcrowded and prisoners often share cells it would be difficult in the short term to ensure that homosexual acts did not take place. Education programmes have, however, resulted in an appreciable decline in the incidence of sexually transmitted diseases among homosexual men.⁵ Although a more heterogeneous group than the gay community, and although the effects may be difficult to assess, prisoners should be offered similar education

programmes. Avoiding risky activities should be encouraged, but if homosexual activity is going to occur then safer sexual practices should be encouraged. Trained people from the community should be asked to help in such programmes.

Issuing condoms to prisoners is controversial but may in the short term be the only helpful means of reducing the risk of the sexual transmission of HIV. The virus does not pass through the intact membrane of latex condoms, and their use provides substantial but not complete protection against infection (R Detels *et al*, fourth international symposium on AIDS, Stockholm, 1988).

In Britain a homosexual act in private is not an offence provided that both parties have consented and are 21 or older. The act is not considered as private if more than two persons are present, and a prison cell is not regarded as a place of privacy. Issuing condoms is thus seen by some as condoning an illegal activity, but their anxiety must be balanced against the possible benefits of distributing condoms. For example, are all prisoners to be issued with condoms? If not from whom

does the prisoner obtain them and how may confidentiality be maintained?

The only alternative to issuing condoms is, however, the enforced isolation and close supervision of prisoners during social intermingling. Minimising the work of HIV transmission among prisoners is clearly an issue for prison authorities to address urgently.

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- 1 Prison Reform Trust. *HIV, AIDS and prisons*. London: Prison Reform Trust, 1988.
- 2 Scottish Home and Health Department. *Prison statistics (Scotland) 1986 No 6*. Edinburgh: SHHD, 1986.
- 3 World Health Organisation. WHO consultation on prevention and control of AIDS in prisons. *Lancet* 1987;ii:1263-4.
- 4 Harding TW. AIDS in prison. *Lancet* 1987;ii:1260-3.
- 5 Carne CA, Weller IVD, Johnson AM, *et al*. Prevalence of antibodies to human immunodeficiency virus, gonorrhoea rates and changed sexual behaviour in homosexual men in London. *Lancet* 1987;ii:656-8.

Palliative medicine

A new specialty

After 21 years of pioneering work by Dame Cicely Saunders the hospice movement and other specialist agencies for dealing with the dying have come of age. In November 1987 the Royal College of Physicians recognised terminal care as a new subspecialty of general internal medicine and called it palliative medicine. Subsequently the Joint Committee on Higher Medical Training has approved a training programme for senior registrars in palliative medicine.

Two decades ago there were a handful of charitable hospices requiring few doctors and no consultants. In 1988 there are 93 independently managed hospices with 2349 beds and 31 units managed by the NHS with 476 beds. Six more units are planned to open in the next 12 months. In addition, there are 231 domiciliary teams that bring the skills of palliative medicine into the community and 21 teams providing similar services within hospitals. Unfortunately, financial expedience may render the word "team" a misnomer: sometimes it is only one or two nurses without medical, ancillary, or secretarial support. The initiatives of the royal college should eventually enable nurses to obtain the medical support they need and to form the interdisciplinary team that is essential for effective hospice medicine.¹

These developments have been accompanied by two other initiatives. First, the Department of Health and Social Security asked all health authorities in February 1987 to review their services for patients who are terminally ill.² Many responded by forming a terminal care planning team to identify gaps in existing services, propose suitable remedies, and plan developments. Already this has led to a demand for more doctors trained in palliative medicine.

The second initiative was the formation of the Association of Palliative Care and Hospice Doctors of Great Britain and Ireland. After fewer than three years the association has 205 members, including 66 full time consultants or medical directors and 22 junior members in various training posts. Most of the remainder are radiotherapists, medical oncologists, physicians, surgeons, anaesthetists, and general practitioners working in or collaborating with specialist terminal care services. Eleven members are in full or part time

academic posts in palliative medicine, nine of which are funded by Cancer Relief for up to five years. In addition to holding regular scientific meetings the association has subcommittees on education, training and manpower, and ethics and research. There is an active junior members' forum. Although not part of the association, there is also the *Journal of Palliative Medicine*.

The proposed training programmes will either train senior registrars for up to four years to become full time consultants in a hospice or a hospice team or provide up to one year's structured experience for those entering another specialty—for example, radiotherapy or medical oncology. For the first five years the scheme will be flexible with entrants having a broadly based medical background leading to membership of the Royal College of Physicians or other appropriate qualification, which will include membership of the Royal College of General Practitioners to ensure that senior posts remain open to family doctors.

The equivalent of four to six new consultant posts are expected each year for the next few years so any bottleneck seems unlikely in the short term even if blocked senior registrars from other disciplines pursue a career in palliative medicine. When the log jam in other cancer services is relieved by creating more consultant posts and by the proposed district cancer physicians³ the ideal candidates for the future must be those who, after completing general professional training, wish to make palliative medicine their specialty.

Manpower planning for the NHS services will be provided by the Joint Planning Advisory Committee: no such planning mechanism yet exists for the independent hospices and teams. The hospice movement must face this challenge by controlling itself through the Association of Palliative Care and Hospice Doctors of Great Britain and Ireland and by persuading the Joint Planning Advisory Committee to include the independent services in its deliberations.

Some may argue that this emphasis on specialist training will detract from the work, training opportunities, and status of the part timer or generalist in the hospital and the