The incidence, prevention and therapy of hepatitis associated with blood product therapy is part of the research programme. The immunological response to blood product therapy and the way in which this is affected by HIV and hepatitis infection is also being studied. There is a team of scientists working on platelets, particularly the platelet membrane, and this will eventually improve the treatment of patients with congenital platelet disorders.

Education

In 1974 the centre was designated as an International Haemophilia Training Centre to which staff from centres worldwide could be sent on training fellowships. In addition we are frequently visited informally by staff from other centres, and this is reflected in our visitors' book 1987-8, with entries from India, China, New Zealand, Poland, Egypt, USA, Australia and Ireland.

As part of a medical school we regularly teach medical students about all aspects of haemophilia care. Every six months there is a two-week course for doctors who are near the end of their training as haematologists and who will soon become consultants. This has a particular emphasis on the laboratory aspects of haemophilia care.

The future

Our aims for the future are in the improvement of patient care – especially in the evaluation of safe blood products; in the provision of efficient and accurate genetic counselling; in the development and utilisation of effective laboratory tests; and in the continuing development of education and research.

Haemophilia and Haemostasis Centre
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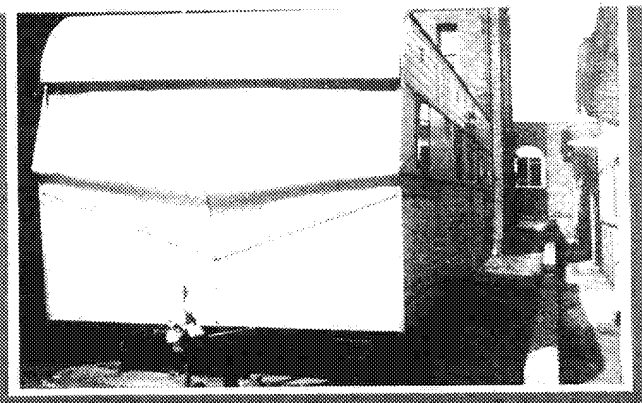
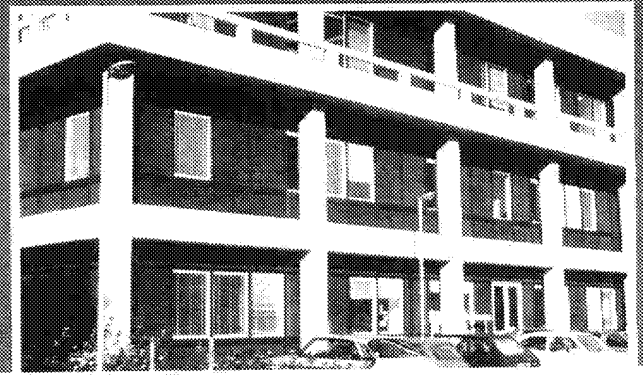
1987



HAEMOPHILIA CENTRE AND HAEMOSTASIS UNIT



Christine A Lee MA MRCP MRCPath

The Katharine Dormandy Haemophilia Centre at the Royal Free Hospital, Hampstead, opened ten years ago, was made possible by a generous financial contribution from Mr Laurence Knight. In one way, the opening was a sad occasion because the director, Katharine Dormandy, who had the inspiration for the new centre, died shortly after. However, she did see in concrete terms the culmination of many years' work and the beginning of a new era in haemophilia care.

The beginnings

The Royal Free Hospital Haemophilia Centre started in 1964 with five patients and the specific objective of providing a good treatment service for haemophiliacs living locally. The space for the developing centre was very cramped. Outpatients were given their infusions at the end of one of the wards of Lawn Road, an old fever hospital which occupied part of the site of the modern Royal Free Hospital. There was a small amount of laboratory space for coagulation work, and the office for the consultant and research assistant was a partitioned end of a staff coffee room.

A significant landmark was the donation of a large caravan by the Haemophilia Society in 1965. This provided accommodation for the two research assistants who were studying the medical and schooling problems of young haemophiliacs in south-east England. This caravan was to remain a prominent land-mark at Lawn Road until demolition in 1973.

The World Federation of Haemophilia

As a consequence of the registered patients seeking advice about treatment facilities when travelling abroad, together with reciprocal requests from others, the centre became an agency to whom patients and staff telephoned or wrote from all over the world. In 1968 the World Federation of Hemophilia brought about a scheme for correlating such information, and Dr. Dormandy's research assistant, Mrs Peggy Britten, became its regional secretary.

Doctors, technicians and medical students soon became regular visitors at the caravan-office to discuss the haemophilia problems in their home towns and to see the Royal Free Hospital Haemophilia Centre.

Growth of the Centre

By 1970, the centre had 180 patients. With this fast growth an outpatient treatment unit became a necessity and this time the Haemophilia Society provided for a prefabricated extension on the verandah of ward 7 at Lawn Road. At that time treatment was predominantly on an outpatient basis, with infusions of fresh frozen plasma.

Cryoprecipitate and home treatment

It was in 1970 that the centre took part in the trial production and clinical evaluation of cryoprecipitate, and its production was soon taken over by the North London Blood Transfusion Centre. In 1971 the first patients were begun on home treatment with cryoprecipitate, soon to become a routine in the centre and later, other parts of the country. This, of course, provided a great freedom for patients, who no longer needed to attend the centre each time they had a bleed.

By 1972 there were 220 patients with inherited bleeding disorders registered at the centre. These included 132 with haemophilia A (factor VIII deficiency), 28 with haemophilia B (factor IX deficiency), 52 with von Willebrand's disease, 1 with factor VII deficiency and 7 with factor XI deficiency.

The new Katharine Dormandy Haemophilia Centre

The present director, Dr P B A Kernoff, together with Dr E G D Tuddenham, who has now left the Royal Free, joined the centre in 1978.

At that time treatment with freeze-dried clotting factor concentrate was gradually replacing treatment with cryoprecipitate, and most severely affected haemophiliacs were becoming established on home treatment.

During the past ten years there has been an enormous expansion of the centre, reflected in the number of patients – 981 – registered for the period 1987–88 (all patient data is entered on to the centre's computer when he or she is first registered): 424 with haemophilia A, 95 with haemophilia B, 204 with von Willebrand's disease, 68 with factor XI deficiency and 142 with congenital platelet disorders. These patients attend from a wide variety of areas : 39% from the North West Thames region ; 30% from the North East Thames region ; 21% from other National Health Service regions and 7% from abroad.

Patients of all ages are registered with the centre and almost one hundred are less than ten years old. We are also a haemostasis unit and of 550 new patients seen in 1987, about half had problems of clotting.

This increase in patient numbers has required more staff. The staff of the Centre now includes: the medical director, Dr P B A Kernoff; a consultant haematologist, myself; an associate specialist, Dr. Eleanor Goldman; the staff for the routine laboratory and a small genetics laboratory, a senior scientist, Dr Ron Hutton; a research development scientist; a research team; three nursing sisters including Clinical Nurse Manager Patricia Lilley; a data processing officer, an office manager, secretaries and a reception co-ordinator.

Professor Roger Hardisty, who recently retired from Great Ormond Street Hospital, has joined us as Emeritus Professor. Three members of our current staff, Mr David Bone, Senior Chief MLSO, Mrs Riva Miller, the medical social worker, and Mrs Peggy Britten, our volunteer data processor, worked at the haemophilia centre at Lawn Road hospital.

The review system

All patients are regularly reviewed. Those with severe haemophilia are seen every six months, and those with mild haemophilia and other inherited disorders of coagulation are seen annually. The reviews provide an opportunity for regular assessment of the haemophilia, general medical health, dental care, the need for hepatitis B vaccination and social aspects of care.

All patients complete home treatment forms and information from these is entered on to the computer. This enables a print-out of annual or six-monthly treatment usage to be provided at each review. It also provides precise information about the number of bleeds into each joint and hence, target joints.

A monthly orthopaedic clinic is held jointly with the consultant orthopaedic surgeon, Mr J C A Madgwick. These clinics are also attended by the physiotherapist, medical social worker and regional liaison sister together with a sister and doctor from within the Haemophilia Centre. Patients can self-refer to these clinics, but more often problems are found at the regular review, necessitating referral to other departments.

Orthopaedic surgery

Patients are admitted to an orthopaedic ward for operative procedures. There is close co-operation between the orthopaedic team and the Haemophilia Centre. All factor concentrates are administered by the sisters from the centre and there is careful monitoring of the levels by the routine laboratory. The surgeon will not operate unless the pre-operative factor level is known. The orthopaedic nursing is carried out by the ward nurses and there is close liaison with the laboratory when specialised nursing procedures or physiotherapy are required.

Genetic counselling

This has always been an integral part of the clinical work, but more recently, with the advent of genetic probes enabling DNA analysis, this work has expanded and there is now a small genetic laboratory devoted entirely to it. Although this is primarily an NHS service, the Haemophilia Society provides substantial financial support.

Chorionic villus sampling is performed at the Royal Free Hospital to ascertain whether a fetus has the haemophilia genotype. An unusual aspect of this work is the genetic counselling for families with factor XI deficiency. The Royal Free Hospital is situated in Hampstead district, with its large Jewish population, and thus a relatively high number of patients with factor XI deficiency are registered at our centre. There is the additional problem of the inadvisability of home circumcision in such patients.

Fetal blood sampling for factor VIII or IX levels is performed at King's College Hospital.

The impact of HIV

The comprehensive care given by our centre has provided a natural framework to help cope with the devastating problem of HIV infection. All patients who are anti-HIV positive are seen for at least six-monthly review: 90% of these patients have severe haemophilia. These reviews provide an opportunity not only for a full medical assessment, but to address HIV-related issues and to ensure that patients have full information about their situation.

Now, with the advent of specific anti-viral therapy, particularly AZT, patients are seen more frequently, on a two-weekly basis, to monitor blood counts. A number of patients are receiving nebulised pentamidine as a prophylaxis against pneumocystis pneumonia. This is administered by one of the sisters to outpatients in the haemophilia centre, and we hope to move on to home therapy.

Patients who are ill with HIV infection and need inpatient care are admitted to one of our beds on a general medical ward. This ward also has beds for patients with chest disease and there may be in-patients with AIDS from other causes. The staff of this ward are thus developing increasing expertise in the care of AIDS. There is always close liaison with the staff of the haemophilia centre.

Counselling

Our medical social worker, Mrs Riva Miller, has always applied the techniques of family therapy and is one of two trained family therapists (the other being Dr. Eleanor Goldman) within the Haemophilia Centre. Patients are seen by her at the time of reviews and are encouraged to bring relatives or close friends to such interviews. Sometimes, if there is a specific problem, a family interview will be especially set up. This way of working has naturally extended to HIV-related problems and it has been used to help address dreaded issues.

An integral part of the counselling is the organisation of groups. These meet once or twice yearly, for about two hours on a week day evening. We have carefully structured groups for parents of younger children, adolescents, wives, adults and, more recently, for HIV-positive patients. They provoke hard and sometimes painful discussion and are certainly not a forum for gossip. We also have a staff support group, held monthly, mainly for 'hands-on' carers, particularly the nursing sisters who are very heavily involved in all aspects of patient care

The routine laboratory

The routine laboratory provides the essential facility for the diagnosis of new patients and the monitoring of registered patients who are receiving treatment. There is also an active development team for the introduction of new laboratory tests. The National External Quality Assurance Scheme headquarters has been recently sited within the Haemophilia Centre and this provides for a national quality control of laboratory tests.

Research

The Haemophilia Centre has always been closely involved in the development of safe blood products. Treatment of patients with inhibitors using porcine factor VIII was pioneered at the Royal Free Centre. The purification of factor VIII by Dr Tuddenham and his co-workers led the way to the production of genetically engineered factor VIII, and evaluation has recently started in the patients in our centre.