APPENDIX A1

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EDITORIALE

Guidelines For The Organisation Of A Haemophilia Centre

The first Haemophilia Centres were set up in the United Kingdom in 1954. Originally they were laboratory based, the chief function of their staff being to diagnose with accurancy patients referred to them. As methods of treatment have changed with the introduction of newer and more potent blood products, the role of the Centres has become much more broadly based and their staff are nowdays responsible for the everyday care of patients with bleeding disorders at every age.

Within the United Kingdom there are now over 100 Centres, the majority of which treat a few patients from the surrounding area on a day-to-day basis. In addition to these smaller centres there are a number of larger, so-called Reference Centres where patients with complications can be referred. Thus the whole country is covered and no patient need go without expert guidance for geographical reasons.

With advances in treatment has come increasing emphasis on the need for expert counselling of families. This aspect of haemophilia care is sometimes given low priority, but it is vital to the health of both the patient and the family. Counselling is dependent on experience of many patients with haemophilia. It cannot be performed, whether in the generic or in the genetic sense, if the doctor or paramedical worker involved has not had this experience. An example of poor counselling occurs when a child with haemophilia but no other problems is referred to a special school for the physically handicapped when he should go to normal school.

Counselling requires privacy, time and patience. It needs a working knowledge of severity, prognosis and general, social and environmental medicine.

Families need advice about education and careers, and as children grow they need to set reasonable goals in acti-

vities and sports. Although much of this counselling can take place within the formality of a clinic session, parents and patients should always be incouraged to seek advice informally if they wish. Some Centres run special group sessions, for instance for the mothers of young children or for adolescents.

Whatever the arrangements for counselling it must in the first instance be placed on very accurate diagnosis. Not only do the haemophilias need to be differentiated, but haemophilia A must be distinguished from von Willebrand's disease and an accurate level of the clotting factor involved must be known in order to guide consultation about prognosis. It is especially important that diagnosis is early in a child's life. When there is a family history, cord blood diagnosis should be available. Some families will need advice about intra-uterine diagnosis by fetoscopy.

Factor assay for diagnosis and the monitorng of treatment demands skilled laboratory staff and an everyday reliance on careful quality control. This can be achieved by the use of national or international standards, by comparative studies between laboratories and by a high frequency of investigation.

Because laboratory screening is expensive, laboratory resources for a geographical reason might best be concentrated in one Centre. Once diagnosis is made the patient or parents should be given a card with the details of the disorder, and should know and where to find help at any time of the day or night.

There is a growing tendency to start prophylactic therapy very early in life for the severely affected haemophiliac. It is not yet our practice in Newcastle to begin home therapy until the child has started formal schooling. By this time the parents know how to recognise which bleeds to treat, have overcome their fear of the disorder, and are happy with the child starting normal school. The doctor also has knowledge of the clinical severity of the child's disorder. Veins easily accessible and parents soon become expert in giving the necessary concentrate.

The organisation both on-demand therapy at home and prophylaxis is the responsibility of the Centre. In the United Kingdom patients can only get their factor concentrate from a Centre; it is not customary for a family doctor to prescribe a blood product, as is the case in Italy. The treatment link with the Centre provides the foundation for follow-up of both the patient's general health and development and of his haemophilia. We run clinics for adults at least once a year and for children six-monthly. More frequent appointments can be given for special reasons, for instance the use of prophylaxis.

The figure shows the organisation in Newcastle in diagrammatic form (Figure 1 here). The Centre is staffed by a core team who work either full-time or part-time with haemophilia and related disorders.

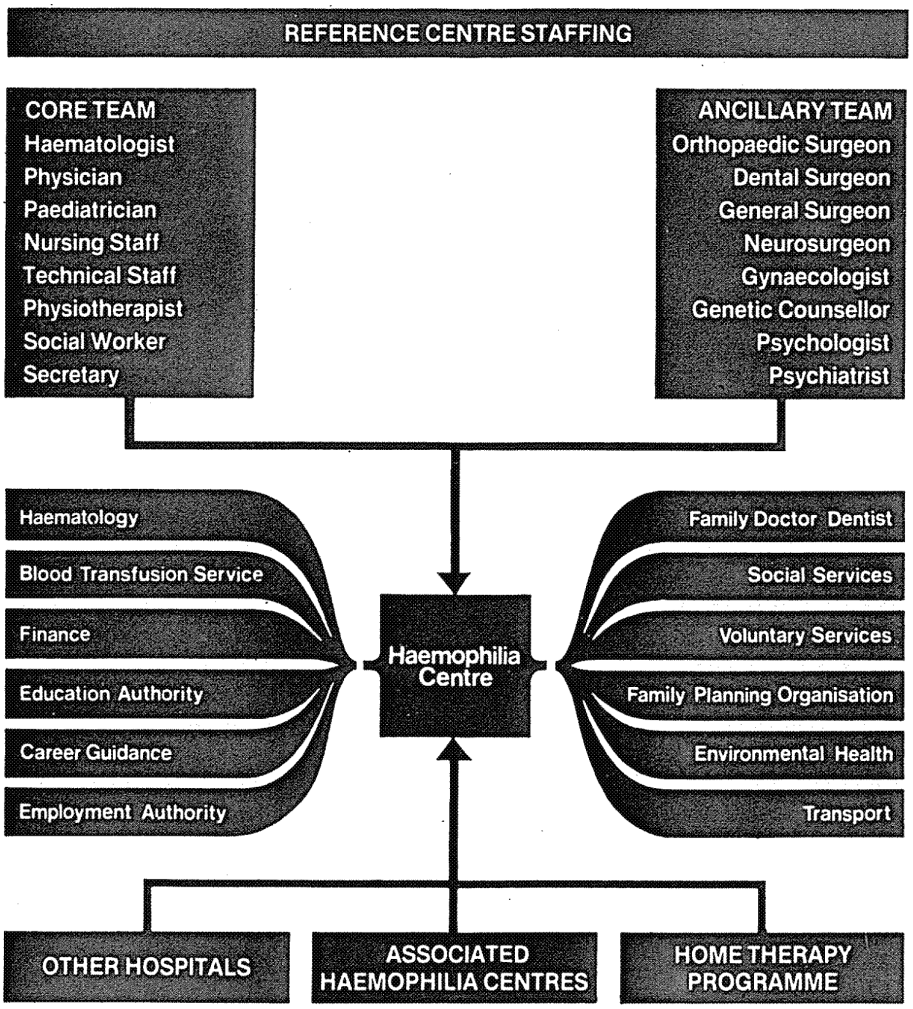

Fig. 1 Reproduced from Jones P. Haemophilia Management: a Physician's Guide to the Tratment of Haemophilia. Cambridge Transart, 1979.

There is a good communication between this team and ancillary members of staff who know enough about haemophilia not be frightened by it, and who can be called in at any time to give expert advice and treatment.

The centre is also linked to a number of other organisations varying from the Haematology Laboratory and Blood Transfusion Service to institutions concerned with

education and employment and organisations like the Social Services and family doctor schemes.

With the agreement of colleagues within our geographical region, which has a population of 3,3 million, the home therapy programme is run from the Newcastle Centre and all patients are followed up there by members of the core team. Lines of communication between the Reference and Associate Centres and between the Reference Centre and District General Hospitals - where one or two patients might be treated - must be established clearly or complications could easily arise; for instance a haemophilic receiving physiotherapy can be injured if the therapist is not aware of limitations imposed by bleeding.

It is particualry important to have regular meetings of the core team. We do this at weekly intervals. These meetings allow members of staff to voice their opinions about individual patients and about the organisation of the Centre in an informal atmosphere. Support can be given to individual members of staff, and manipulation by particular patients discussed.

We make it a rule in our home therapy programme not to supply blood products unless a very careful record is kept of their use by individual patients. In this way we have detailed knowledge of all bleeds and their response to treatment when the patient comes for follow-up. We have found it extremely useful to keep a calendar log of bleeds, rather than a written diary of episodies. Target joints can be spotted immediately and the effect of changes in treatment seen easily.

Finally, of course, every Haemophilia Centre must provide 24-hour cover. In the United kingdom, under the terms of the National health Service, all treatment is free and Haemophilic patients are given immediate entry to hospital, simply phoning up for an ambulance if they are in trouble. Patients with haemophilia should not be treated in Casualty Departments by inexperienced staff. Instead, a designated ward or wards for children and adults should be known, and all treatment given outside outpatient clinic hours should be concenrated on these wards.

Of especial importance in the management of a rare disorder like haemophilia is the pooling of experience by the different Centres.

Figures for the United Kingdom as a whole are kept by the Oxford Haemophilia Centre and discussed anually between the Directors. Such discussion provides for peer review and a certain amount of audit.

Peter Jones, MD, FRCP, DCH
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SUGGESTED FURTHER READING:

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