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Figures and photographs have been omitted; fu l proceedings to be published in 1977 by the British Haemophilia Society.

The organisation and role of the Haemophilia Centre in the comprehensive care of the haemophiliac

Members of staff, Newcastle Haemophilia Centre, England

Introduction

Dr Peter Jones

Director, Newcastle Haemophilia Centre

Mr President, Ladies and Gentleman,

It gives me great pleasure to welcome you to this session of the 3rd European Congress of the World Federation of Hemophilia. In this session we are to explore the organisation and role of the Haemophilia Centre in the comprehensive care of the haemophiliac. In doing so we will inevitably touch on many of the topics to be considered throughout this Congress, and will attempt to set the scene for a fruitful discussion on the ways in which, in the light of our present scientific knowledge, the care of the haemophiliac can progress.

In the past decade we, as clinicians, have been handed new and powerful therapeutic weapons by the internationally based research workers referred to by Professor Ingram this morning. As doctors it is our duty to use these weapons wisely to the fullest advantage and hopefully never to the detriment of our patients. Unlike most doctors, except those working in those disciplines which demand human organs for the purpose of transplantation, we have also to be constantly aware that the successful application of haemophilia research is at present totally dependent on the adequate supply of a human resource. Without the blood donor, whether voluntary or paid, the Haemophilia Centre would be a sad place in which to work.

But, given the excellence of the blood products available to us, just

how much of a role in the life of the haemophiliac and his family should the Centre play? Should mature adult haemophiliacs be any more reliant on the medical profession than, say, their diabetic counterparts? Has the time been reached when haemophilia care should be primarily the responsibility of the family doctor rather than the hospital? Does the organisation of a 'comprehensive care system' encourage, rather than discourage, dependency? Should the haemophiliac expect complete, immediate and financially protected access to costly medical and social services or has society the right to expect him to restrict his family, if not his own demands, on the Welfare State?

When the Medical Research Council established the first Haemophilia Centres in the United Kingdom in 1955 many of these questions were academic. The major, and often the only role of the Centres was to attempt to diagnose the haemorrhagic disorders and to manage patients as best they could within the strictures of treatment then available. Although this role of diagnosis remains a fundamental one advances in therapy require that the staff of the present day Centre are conversant with the many complex clinical and social problems that haemophilia presents. This change in emphasis, from laboratory to consulting room and from consulting room to the patient's home and environment, is reflected in the recently issued United Kingdom Department of Health and Social Security Health Circular (HC(76)4), which will eventually be available as a memorandum from Her Majesty's Stationery Office.

This Circular sets out new and strict criteria for the care of haemophilia within a tri-partite structure of Haemophilia Reference Centres, Haemophilia Centres and Associate Haemophilia Centres. It thus recognises both the need to concentrate highly sophisticated and expensive resources and therapy in major Centres whilst continuing to provide the best possible treatment and advice near to the homes of affected families.

In order to perform the three fundamental functions of haemophilia care — diagnosis, treatment and advice — the modern Haemophilia Reference Centre needs a team of people, most working part-time in the field, but all with a fundamental knowledge of the hereditary bleeding disorders (Figure 1) .

I will not dwell on this list (many of the people on it will be addressing you shortly) except to point out how extensive the resources of modern care need to be. In addition to constant attention to communication within this team the active Centre must also maintain links with a host of outside agencies (Figure 2). It must also have the closest communication with the Blood Transfusion Service, and with associated Centres and other hospitals (Figure 3). In our Region a home therapy programme is at present also based on the Reference Centre.

What the figures do not show is the constant input of information from other Centres and from the medical literature on all aspects of haemophilia care. Nor does it indicate the purely local educational and research aspects which inevitably become part of the co-operative effort in any major Centre.

Representing this latter role is Mrs Susan Lewis, a research assistant who has just completed a three year survey of the distribution and problems of haemophilia and related disorders in Northern England. The Survey was conducted with the generous support of our hosts this afternoon.

JONES

Key FIGURE 2

RHA	Regional Health Authority
AHA	Area Health Authority
DHSS	Department of Health and Social Security
DRO	Disablement Resettlement Officer

1. Mrs Susan Lewis

THE NEWCASTLE HAEMOPHILIA SURVEY

In the United Kingdom today there are 6 major Reference Centres and ~~some~~ over 50 associated Centres which treat patients with haemophilia and related bleeding disorders (Figure 1).

The Newcastle Reference Centre is responsible for the care of haemophiliacs in Northern England, a geographical area stretching from North Yorkshire to the Scottish border and from the industrial east coast to the Lake District and as far as Barrow in Furness in the west (Figure 2). The population of the region is 3.3 million.

Although haemophilia care within the region was established in the 1950's by Dr T H Boon in Newcastle and Dr A Inglis in Carlisle, the new purpose built Centre was only finally established during 1974 and its opening coincided with our regional Survey — a project which was financed by the British Haemophilia Society and lasted for a period of three years.

The purpose of the Survey was three-fold:

— to identify and to evaluate the state of each patient;

— to create a picture of past overall care and treatment throughout the region;

— to act as a guide for those creating a new system of care.

From these primary aims came three further functions:

— specific problems raised in the course of the personal interviews could be referred back to the Centre;

— haemophiliacs and their families could be told about the new Centre and the concept of overall care;

— family histories could be checked and carriers identified.

As a result of work using the Hospital Activity Analysis diagnostic index of the Regional Health Authority, records of regional consultants

in haematology and pathology and of family doctors, and the home interviews of families the following Registers were compiled:

1. Patients known to have a bleeding disorder (258)
2. Obligatory carriers of a bleeding disorder (105)
3. Potential carriers and patients needing diagnostic reassessment (288)

The Survey involved the visiting of each patient with a known diagnosis of bleeding disorder at home with a questionnaire; families were asked by letter for their permission to be included in the personal survey before these visits were arranged (one refusal). The questionnaire incorporated the following topics:

— family and family planning (including a complete family history;)
— housing and environment
— transport
— finance
— education
— employment
— hobbies
— medical aspects including an enquiry into pain and the patients' requirements for better treatment.

The questionnaire was therefore designed to provide as complete a picture as possible of each patient's life.

In all a total of 200 of the 258 patients with a known diagnosis were interviewed (the differential diagnosis is shown in Figure 3). This total included virtually all the known severely affected families. An additional 15 patients were contacted but no visit was made; nor were the remaining 43 patients visited as our information indicated a very mild disorder and it was thought that the questionnaire would provoke undue anxiety. All these remaining patients were invited to

attend the Centre for reassessment at a later date. Figure 4 shows the regional distribution of our patients; the distribution is in keeping with that of the normal population.

Within the context of this paper it is not possible to enumerate all the ways in which this Survey helped, and is continuing to help, the Centre staff. However one further aspect is well worth mentioning this afternoon — all the families were asked for their permission for names and addresses to be sent to the local branch of the Haemophilia Society, itself undergoing reorganisation. No-one refused.

The data took two years to collect and was then evaluated with the aid of the Regional Health Authority Statistics Department (headed by Mr Angus McNay) using a computer. Evaluation was based on

— diagnosis and severity
— whether patients were under or over 16 years of age at the time of interview
— whether or not a family history was known.

Reference to some of the results obtained will be made by other speakers, and it is hoped that full publication will eventually be possible.

With the agreement of the British Haemophilia Society copies of the questionnaire used in the Survey may be obtained by contacting Mrs Lewis at the Haemophilia Centre, Royal Victoria Infirmary, Newcastle upon Tyne NE1 4LP, England.

Chairman: Mr Alan Oxley is Chief Technician in the coagulation laboratory of the Department of Hematology of which the Newcastle Centre is a part. He became a member of the team in 1966 when, after training in Oxford and later in Edinburgh, he was responsible for setting up the assay systems we use today.

2. Alan Oxley

THE ROLE OF THE LABORATORY

The role of the Haemophilia Centre laboratory may be discussed under a number of sub-headings, the major divisions being:

- diagnosis
- detection of factor antibodies
- monitoring of replacement therapy and
- carrier detection

In addition the Centre laboratory often serves as a general coagulation laboratory dealing with acute haemostatic disorders and with anticoagulant control.

As Mrs Lewis has already shown, the region served by the Newcastle Reference Centre has a total of 258 patients with a diagnosis of bleeding disorders. Figure 1 shows the breakdown into differential diagnosis for these patients. ~~It also shows that~~ As a result of the Survey, we know of a further 288 patients with a provisional but not yet firmly established diagnosis of bleeding disorder. These are patients who must be retested; they were inherited from the time when laboratory tests were more fallible than they are today, many of them being classified as "easy bruisers". Even if these patients are still found to be haemostatically "normal" by repeat laboratory testing their histories should be respected and extra caution employed when they require surgery.

Concentrating on patients with a deficiency of Factor VIII we find that severity can be graded according to the amount of circulating active Factor VIII present in the patient's plasma. In Figure 1 those patients with a diagnosis of Factor VIII deficiency (classical

haemophilia) have been classified as being severely affected (in our practice < 1%VIII), moderately affected (1-5%) or mildly affected (> 5%). So out of a total of 258 definitive diagnoses we have 178 affected by a deficiency of active factor VIII of whom 50% are in the severely affected category. These are the patients who will require the most intensive treatment and therefore take up most laboratory time.

In order to report with certainty the actual amount of factor VIII present it is necessary to have an accurate assay system. The method we have chosen is a two-stage technique developed by Biggs and McFarlane which is based on the thromboplastin generation test, and in which, by comparing the patient's plasma with a standard pool an actual percentage of factor VIII activity can be estimated. Since we are working in narrow limits at low concentrations it is essential that a quality control programme be incorporated into the assay system. The first priority is for a "normal" control pool. This can be achieved by using a constant selection of donors whose VIII activity has regularly been assayed; fortunately there are always people in the laboratory who can be persuaded that they are in fact "volunteers". The normal range for factor VIII is between 50 - 200% and the greater the number of donors the more likely the pool is to contain the mean of 100% factor VIII activity. The great assumption made when performing assays is that the pool actually has an activity of 100%. It is perhaps justifiable under the circumstances previously stated, but nonetheless remains an assumption.

In order to justify the assumption even further the use of freeze dried standards is recommended. Some of these are available from commercial organisations and come with a stated assay value. The products are stable when in the freeze-dried state, but once

reconstitued are generally only stable for the working day when kept on ice and they cannot be deep-frozen. They are incorporated into the assay system and are of use as a check on the normal pool used for the standard curve.

There are also a British Standard and an International Standard for factor VIII. Before release the standards have been assayed by selected laboratories on numerous occasions and a value given after analysis of the results obtained. Without these standards to gauge the accuracy of an assay results could not be issued with such confidence.

The other inbuilt control is regular use of the assay system. Accuracy increases with frequency of use. A two-stage system which is used daily will give greater accuracy than one which is used only infrequently. Familiarity with the technique leads to recognition of malfunction.

However, the most important "reagent", ~~if you like,~~ in the assay system is a haemophiliac with 0% factor VIII who is willing to give up a few millilitres of blood so that the deficient plasma necessary for the assessment of factor VIII levels in others is available. Without this plasma we would have no assay system.

Although work involving haemophiliacs occupies approximately 60% of the total laboratory time other services are provided. The control of patients on oral anticoagulants, for instance, involved over 11,000 tests being carried out in the last year in Newcastle. These were divided more or less evenly between in-patients and out-patients but involved three members of staff devoting 12 hours per week to this one particular aspect. We also tested 7000 patients over the same period for a variety of reasons, (i.e. control of heparin therapy,

pre-operative screening to assess haemostatic status and diagnosis of possible bleeding disorder). Of these 7000 patients, 300 were seen at a specially devised clinic where complete personal and family histories are taken and the full battery of coagulation tests can be brought to bear in order to establish the exact diagnosis. At this and the Haemophilia Review Clinic the technical staff are in direct contact with the patients themselves; people are no longer just names on request forms. Although a close personal relationship may not be built up at least the technical staff meet the patient; by doing so they can better appreciate the need for accurate diagnosis and gain an insight into the problems the patients may face.

Oneof these problems may be lack of response to an infused product due to the presence of an antibody to factor VIII. The presence of an antibody presents difficulties in treatment but does not prevent a patient being treated. Neither will it automatically result in the removal of a patient from the home therapy programme. Although the titre (or strength) of the antibody will probably rise after treatment some patients still appear to gain relief from bleeding by continuing intermittent use of a factor VIII blood product. At the present time we have 12 patients with antibodies, 2 of whom (originally with low titre antibodies) are on home treatment. In one of these, far from having an anamnestic response to infusion, the antibody has decreased in strength until it is no longer detectable. On the other hand, another patient with antibodies received a total of 108,670 units of factor VIII over a period of ten days, before bleeding stopped. The titre of this antibody was rather high but was eventually swamped.

CARRIER STATUS:

Carrier detection should be approached with the greatest caution. The consequences of informing a woman that she is a carrier will

of course be disturbing. Even more disturbing would be to tell her she is not a carrier and then deliver an affected baby. In early studies carrier detection relied solely on the biological assay of factor VIII clotting activity (VIII:C). The average VIII:C level of women who were obligatory carriers (mothers of more than one haemophilic son or daughters of a known haemophiliac) was found to be 40% of normal average. The range was something like 22% to over 100% compared with the normal range of 50-200%, so not surprisingly the detection rate of genetically proven carriers was as low as 25% or less (Merskey, 1951, Rappaport 1960, Veltkamp, 1968).

More recently it was found by Bennet and ~~Antigen~~ Heuhens and Zimmerman and Ratnoff that an antibody against factor VIII raised in rabbits could detect a factor VIII like material in the plasma (VIII R:AG) which was useful in discriminating between carriers and non-carriers. This antigen when compared with the VIII:C helped in determining carrier status. Even when the VIII:C level was depressed, the factor VIII R:AG was normal, and when expressed as a ratio of VIII:C to VIII A:AG, carriers were found to have a lower ratio than non-carriers. The difficulty then arose as to where the line should be drawn so that women were not misclassified.

(Figure 2 HERE)

By taking into account VIII:C level alone, ratio of activity to antigen alone, and both level and ratio, there is still an overlap between carriers and non-carriers which works both ways, 16% known non-carriers being wrongly classified and 18% obligatory carriers being classified as non-carriers (Meyer; ~~and~~ Rizza). Therefore one can <u>never</u> tell a woman that she is <u>not</u> a carrier. In accordance with international agreement our present policy is to have three specimens from women anxious to know their carrier status before

even attempting to make a decision. Hopefully this replication will avoid the odd spurious one-off result, and will enable us to determine carrier status with a little more certainty.

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Chairman: Mrs Jean Lovie, who has been a most welcome newcomer to the team in the past year, is a social worker employed by the Local Authority. She works part-time in the Centre, what is left of her time being devoted to the care of her family. Her special interest is with the young haemophilic family.

3. Mrs Jean Lovie

SOCIAL WORK INTERVENTION WITH FAMILIES WITH YOUNG HAEMOPHILIACS

In the course of the Newcastle Haemophilia Survey (1973 - 74) it was found that, of 114 severely affected adults between 16 and 65 years, 33% were unemployed. At that time the regional unemployment figure was 5.1% and the national figure 2.9% These figures clearly indicate the frustrations and lost opportunities for some adults and suggest that there is a need to look at ways of helping the younger haemophiliac and his family with a view to the future. Help with social and emotional problems must be considered as an essential part of a team approach to the child's care which is built round improved medical treatment.

All families may from time to time require help of a practical nature with:

a. REHOUSING. Sixty per cent of severe and 50% of moderate/mild haemophiliacs interviewed in the Newcastle Survey were living in property built before 1919; these figures compare with the national General Household Survey (GHS) figures of 31.0%. Of these living in houses built after 1945 the GHS percentage of 42.0% compares with 18% for severe haemophilia, and 26% for moderate/mild haemophilia. Haemophiliacs tend to live in older properties due to difficulties in obtaining a mortgage and to the prohibitive price of newer properties for those in less secure, less well-paid employment.

b. INFORMATION ABOUT STATE BENEFITS. Help is often required in claiming state allowances, including those available for constant attendance and mobility.

c. LIFE AND HOLIDAY INSURANCE. Close co-operation between Centre staff and insurance brokers or companies results in the granting of policies with little, if any, excess charges for haemophiliacs.

d. TELEPHONE INSTALLATION AND RENTAL. 74% of haemophiliacs known to the Centre now have immediate access to a telephone.

e. INFORMATION ABOUT NON-STATUTORY SOURCES OF FINANCIAL AND OTHER HELP. These include the Central Council for the Disabled, the Joseph Rowntree Trust and of course the Haemophilia Society.

Although problems relating to these aspects of life may be dealt with by a number of other agencies, including the family doctor, the Newcastle Survey established that the haemophiliacs usually relied on hospital staff for help, and for information about their disorder.

In order to determine the degree of social work involvement by the Centre it is important to distinguish between three groups of families.

1. There are those families with social and emotional problems beyond those simply associated with haemophilia e.g., children at risk. The haemophilic child may exacerbate difficulties already present or act as a focal point for deeper family tensions. In the U.K. the needs of these families are best met by referral to their Local Authority Social Services Departments working in conjunction with the Centre Social Workers.

2. There are families who appear to function reasonably well despite the problems of haemophilia. These families are followed up at the six monthly review clinics but left to refer themselves back for help if they wish in the meantime.

3. There are families who have social and emotional difficulties related directly to haemophilia. These families often have a severely affected child. Centre social work should be directed towards this group, Social Workers in associated hospitals seeing families who live some distance

from the Regional Centre.

What is the Social Worker's role in helping this last group of families? It is often possible to anticipate difficulties associated with certain stages of the child's development — social work help should be offered if social or emotional problems appear at these stages. The aim should be to offer short term help to parents at the time of diagnosis,

when the child starts nursery(play)school,

when the child starts formal school,

and when the child starts home therapy.

In addition, if school learning or behaviour problems are reported by teachers or parents, or if it seems that the eight or nine year old child is not well integrated with his peers and family or is not being permitted to take more responsibility for his haemophilia and his treatment, skilled counselling is often necessary.

I would like to briefly discuss social work involvement at each of these stages.

1. Time of diagnosis

Social work follow up at home helps to establish the link between Centre and family. If a visit is delayed for a month after the confirmation of diagnosis, especially where there is no previous family history of haemophilia, parents have time to assimilate some of the medical information they have received. This arrangement also allows time for some of the day-to-day worries about management to emerge. Twenty per cent of the haemophiliacs interviewed in the Survey reported that the management of haemophilia had been a real source of personal conflict between their parents. The chance to express feelings about the diagnosis, and possible guilt reactions, helps parents to adopt a positive approach to the child which other Centre staff are trying to encourage.

2. Admission to Nursery (Play) School or Primary School

For most parents this stage represents the first break in the close parent/child relationship and it is not surprising that, especially when a child has needed extra supervision, parents should be anxious. The policy ofthe Centre is that, preceding admission to school, a letter is sent from the Medical Director outlining the child's medical history and giving recommendations for his management in the light of the continuing improvements in treatment. Nursery school staff sometimes doubt their capacity to cope with the haemophilic child and the Social Worker links Centre, school and family. In the UK the more formal type of Education Department Nursery appears to have advantage over the Social Services Department Playgroups.

3. Home therapy

With the exception of telephone provision, to date is has not been the policy of the Centre to automatically offer social work help to all those started on home therapy. However, as the home therapy programme expands it is becoming clear that help is often needed, particularly during the first year, and the policy is changing. In any case, intervention is appropriate when social factors are temporarily affecting a family's capacity to use home treatment facilities, or when families are temporarily finding it hard to adjust to the home therapy regime after being used to hospital based treatment. In the latter example a joint approach by the social worker and the nursing Sister in charge of the programme is probably best.

4. School Learning or Behaviour Difficulties

By the time a child has been at school for a year or so the accumulative effect of "days off" begins to show in poor reading and writing performance levels. The social worker can help in four different ways -

— by encouraging parents to visit and talk with the school staff.

— by contacting the school to convey the Centre's continuing interest in the child.

— by discussing with the school staff possible remedial teaching.

— by enquiring into the school and home situation in an attempt to determine why difficulties might be arising.

> One boy of six years changed teacher and his work deteriorated. On enquiry it was found that his new teacher, afraid of haemophilia, had moved him to a position at the back of the classroom. If the child complained of pain he was sent to the staffroom to play with an electric train. It was not hard to guess where he had been spending much of his time.

Schools often see the child's needs in physical terms (for example, of how to accommodate a child with a splint or the temporary use of a wheelchair), the haemophiliac being treated as "someone different". Teachers should be encouraged to place emphasis very positively on how the haemophilic boy can contribute to school life in his own way.

4. Social Integration

Enquiry about hobbies, possibly club membership, family activities and interests can often reveal the patterns of behaviour and attitudes that the haemophiliac and his family have adopted to cope with bleeding episodes. The strain of day-to-day care of the affected child and his siblings should be appreciated, and the parents helped to talk out anxieties as a way of helping the child's transition from parental to self responsibility in treatment and in life-style.

Chairman: Mrs Maureen Fearns joined the team in 1973 as the full-time nursing Sister. Known as "Auntie Maureen" by the children she is expert at venepuncture, orthopaedic nursing, running the home therapy programme, making coffee and hiding the sweet tin before the entry of the dentists.

4 Maureen Fearns

THE ROLE OF THE HAEMOPHILIA CENTRE SISTER

My experience in nursing haemophilia has been intermittent over a period of 14 years, during which time I have seen a great change in the role of the nurse. Until recent years treatment for haemophilia was very inadequate, a bleeding episode usually meaning an admission to hospital for bedrest and numerous plasma transfusions. The role of the nurse was then little more than to make the patient's bed and keep him comfortable in a ward full of patients with varying disorders. Little time was available for dealing with the haemophiliac's special needs.

With the inception of the new Newcastle Haemophilia Centre in 1974 I was employed as a full-time Sister to become part of a complete team to care for the haemophiliacs. Since starting the job I have found my role continuously changing and varying. This is a gradual process of getting to know the patients and their relatives more personally. Because they see the same person at every visit they have eventually grown to look upon me as a friend to confide in instead of "that person who puts the needle in".

When a haemophiliac needs treatment for a bleeding episode he can contact me directly by "bleep" and I will have the appropriate ~~IV~~ intravenous (I replacement therapy ready for his arrival at the Centre. This is most important as early treatment reduces pain, cripping and time lost from school or work. After erecting the IV therapy and attending to any additional treatment such as plaster splints, I spend a lot of time talking to the patients about work, family, hobbies and anxieties - often acting as the link with other members of the team, especially the social workers. The physiotherapist and I work closely together as a continuous assessment of joints and muscle-power changes is

needed.

Haemophilia involves the whole family and so relatives and girl friends are encouraged to visit the Centre. Often one knows as much about a prospective fiancee as the patient himself. On many occasions relatives on their own appear for a chat and a cup of coffee. It may be that it is just a social visit but often they pass on invaluable information which will help in the treatment of their haemophilic relative.

Over the past three years the haemophiliac has come to realise that it is only in extreme circumstances that he will be admitted to hsoptital for treatment. This is usually when there is an antibody to FVIII present, or a more serious bleed - for example a head injury or psoas bleed. Adult patients are admitted to one specific medical ward and children to one specific paediatric ward. The nursing staff on these wards are used to looking after the haemophilic patient and are aware of his special needs. Good liaison with ward staff is essential and a quick cup of coffee after the daily ward round facilitates time to discuss problems and exchange information.

All special procedures and treatment are given to the patients on the wards by me. In the past I spent a great deal of time on the wards as there were, on average, six patients in at the same time. Since the home therapy programme started the haemophilic in-patient is noticed more by his absence: most haemophilic patients who are now admitted to hospital are those who are in need of elective surgery. Here my job is to help prepare the patient mentally and physically for operation. For this I take all blood specimens and erect all IV replacement therapy pre - and post-operatively. Having taken the blood specimens I relay the results to the Director so that he can assure the surgeon that haemostasis is adequate for him to commence operation.

The Director or his Deputy and I attend all operations in order to assess the extent of the lesion.

As long stays in hospital are necessary after orthopaedic surgery, one of the most involved parts of post-operative care of the haemophiliac is trying to keep him physically and mentally occupied as we have no occupational therapist. This usually means a trip into town to buy model ships and spending some time talking to the patient each day.

Due to improved laboratory techniques clinics are now held to detect carriers and diagnose new patients with a bleeding disorder. On these days I often spend time talking to patients about haemophilia and offering advice to mothers of newly diagnosed babies. These people often need reassurance and a guide as to what to look for and expect to happen to their son at the different stages of his life.

The majority of times that the haemophiliac needs treatment for a bleeding episode he may only see me and so it is necessary to hold clinics for formal assessment. We aim to see adult severe haemophiliacs annually and children six-monthly. On these occasions I act as the co-ordinator between the patient and consultants, physiotherapist, social worker and dentist and - last but not least - provide coffee or lunch for the patient at the end of a very exhausting visit.

One of the most rewarding parts of my job has been to run a home treatment programme. I say this because it is wonderful to see how it has changed the life of the haemophiliac and his family to that of relatively "normal" people. There are at present 54 patients in the Newcastle region receiving home treatment.

Our suggested criteria for home therapy are as follows

Patients should:

1. be over 6 years of age
2. have suitable veins
3. bleed frequently enough to warrant the training and expense involved.
4. have a stable personality
5. have a capable relative or friend who will be present during injections
6. have no history of chronic drug dependency

In addition the home should contain a telephone and a reliable refrigerator

To start on the programme the patient and relative attend the Haemophilia Centre for two consecutive days during which time they are taught which bleeds to treat, the method of administering treatment and everything else that they may need to know. They are supplied with ten vials of FVIII concentrate and are issued with further supplies from the Centre when they have only two vials left. At this time they have blood specimens taken and a copy is made of their recording book.

Figure [illegible]

On the whole patients adapt well to treating themselves but sometimes problems do arise as they realise that the enormous responsibility of deciding when to treat themselves is theirs. Also, continuous use of veins sometimes causes problems until the patients and relatives become more skilled at the art of venepuncture. Often during the first few months there are frantic 'phone calls from mothers, but fortunately their problems have been only minor ones relieved by the opportunity to talk them out. Equally, I am now used to being telephoned, when off duty, by a triumphant boy who has at last summoned up the courage to inject himself rather than let his parents do it for him!

The aim of home treatment is to prevent damage to joints and therefore reduce pain and crippling. To do this one must work closely with the physiotherapist and have her do regular assessment of joints and muscles. On the whole improvement and reduction in joint damage has been observed but one teenage boy did demonstrate a definite deterioration in an elbow joint whilst on home treatment. This was found to be due to inadequate supervision by the boy's parents and undertreatment for each bleeding episode.

Two very important observations, made from a random selection of 15 patients on our home therapy programme; are shown in Figures 3. and 4.

~~Figures 3 & 4~~

While I perhaps selfishly admit to missing the actual nursing of the haemophilic patient, what greater satisfaction can one ask for than to see these people improve their education, gain promotion in their jobs and actually enjoy a "planned" outing or holiday?

Chairman: Miss Susan Cole is a senior physiotherapist in the Department of Physical Medicine at the Royal Victoria Infirmary, and is attached part-time to the Centre. She joined us in 1973 and has since enjoyed both a World Federation of Hemophilia Travelling Fellowship and, this year, the award of a Winston Churchill Scholarship which will enable her to extend her knowledge of rehabilitation techniques in Canada and the United States.

5. Miss Susan Cole

THE ROLE OF THE PHYSIOTHERAPIST

Today, in spite of early and adequate treatment for the majority of bleeding episodes, the physiotherapist has retained an important role in the management of the patient with haemophilia. One of the major concerns in these patients is the residual damage occurring in joints and soft tissue as a result of repeated bleeding into these structures.

The physiotherapist aims to maintain or restore muscle and joint status and thus minimise the effects of this repeated bleeding. This can be achieved in the following ways (Figure 1)

Assessment

In order to know the range of movement and muscle power to be restored after a bleed, it is vital for the physiotherapist to have a prior record of the measurements when the joint is in a quiescent state. This assessment is of particular importance when a patient starts on home therapy, when it acts as a valuable check on how successfully he is or is not managing his bleeds.

Figure 2 shows how this patient's range of movement improved while on home therapy. As already stated by Sister Fearns, one patient showed a marked deterioration in elbow movement. This was due to lack of parental supervision of home treatment but this was quickly spotted in the assessments and steps taken to remedy the situation (Figure 3).

Instruction

Joint stability depends on the integrity of ligaments, capsule and muscle power. After repeated haemarthroses the ligaments and capsule become lax; it is then vital to have adequate muscle power to help protect and support the joint.

The patient and, in the case of children their parents, must understand the importance of exercise and be instructed in a regular programme of exercises which can easily be performed at home.

A word of caution — responsibility for maintaining exercise must lie with the patient or a rather carefree attitude can result. The threat of withdrawing home therapy or other facilities from the lazy can work wonders!

Splinting

Splinting of a joint can be used either to rest the joint during an acute episode, or to correct deformity or even to prevent bleeding (i.e., a night splint worn for 10 days after a major haemarthrosis often prevents a recurrence). During an acute phase a plaster of paris back splint is applied by the Sister while the patient is in the Centre, with the aim of immobilising the joint and preventing secondary haemorrhage.

Preventive

The ankle splints (Figure 4) were developed with the help of Bolton Bros., Surgical Appliance makers in Newcastle. They provide medial and lateral support whilst allowing movement into dorsiflexion and plantar flexion.

Figure 5 shows the reduction in ankle bleeds in two patients three months before and after having the splints.

Corrective

This splint (Figures 6 and 7) (designed by Mrs Donna Boon in Los Angeles) provides support for weak muscles, and also helps to correct a flexion contracture of about 30°. It is made in a thermoplastic material and can easily be altered as correction occurs.

We also make a posterior splint in this material for those

patients on home therapy with a particularly troublesome joint.

Hospitalisation

Some patients still require hospitalisation, either for a major bleed (a psoas bleed or a patient with antibodies) or for surgery. It had been noticed that patients admitted for bed rest for any reason often had secondary bleeding into other joints or muscles. We therefore commenced a routine programme of bed exercises to all unaffected limbs. Depending on the patient's general condition these exercises can be resisted or unresisted. Correction of poor posture in bed is also vital because this can lead to uneven distribution of weight especially on elbows and shoulders and this encourages bleeding. A firm orthopaedic bed with the affected limb well supported will help to prevent this occurrence by allowing freedom of movement and at the same time not disturbing the affected limb.

Surgery

With effective factor VIII cover many haemophiliacs are now undergoing general surgery as well as preventive and corrective orthopaedic procedures. The physiotherapist must be guided by the medical staff after all forms of surgery as disturbance of the wound before healing is well advanced may precipitate secondary haemorrhage. The main role of the physiotherapist after general surgery (i.e., appendicectomy, vagotomy and pyloroplasty or repair of hernia) is prevention of any chest complications — which may occur after the anaesthetic — and during the first few days that the patient will be in bed. To help this the patient is encouraged to stop smoking and pre-operative chest physiotherapy given.

Initially physiotherapy in post-surgical cases must be carried out after a satisfactory factor VIII level has been achieved.

Physiotherapy after orthopaedic procedures (i.e., synovectomy. arthrodesis or joint replacement) follows the same regime as in a non-haemophilic patient. The physiotherapist must have a knowledge of haemophilia and be aware of the complications that may arise.

Aids to Daily Living

Some of our patients have difficulty with everyday activities. Stiff knees and hips can lead to great difficulty in getting in and out of a chair. This can be overcome by a spring assisted chair which helps to ease the patient in and out.

Having a bath is not much fun if you get in and cannot get out. In this case a bath seat and board can make life very much easier.

Wheelchairs are often a touchy subject. Patients tend to regard them as "the end of everything", but they do allow severely disabled patients access to many things they would otherwise be unable to do or see. Wheelchairs should be regarded as an aid to mobility.

Communication

As many of our patients come from a distance they usually receive out-patient physiotherapy at their local hospital. It is therefore important for the Reference Centre physiotherapist to get to know the physiotherapists at these hospitals; in turn this makes communications very much easier. When a patient is discharged the physiotherapist who assumes responsibility for treatment must be well informed of the patient's history and of the amount of recovery to be expected and aimed for.

Chairman: Mr William Morgan, a Lecturer in Medical Sociology in the Department of Family and Community Medicine of the University of Newcastle upon Tyne, was the first social worker to join the team, as a part-time member, in 1972. In addition to his very full role as a teacher, Bill has found time to help many of the more severely affected of our families. His particular interest is in the care of the young adult.

6 Mr Bill Morgan

SOCIAL WORK WITH YOUNG ADULT HAEMOPHILIACS

We find in Newcastle that care of young people between the ages of 15 and 35 years presents us with many challenges and some very real difficulties. We are intervening in a process of development and this means that present difficulties can rarely be understood except in relation to prior learning. The developing child and his family need to develop coping mechanisms from and early age because, as Fig. 1 shows, all the severe haemophiliacs contacted in the Newcastle Haemophilia Survey had presented before the age of 9 years and almost all were diagnosed before that age. The effects of the disorder are almost immediate and, in terms of the life opportunities presented to haemophiliacs, often disabling. We found that education had often been impaired and ~~Table~~ Figure 2 shows that 40% of our patients who had completed their formal education felt that their disorder had badly affected their education. Not surprisingly employment is also affected and ~~Table~~ Figure 3 shows high rates of unemployment and non-employment for both severe and non-severe haemophiliacs. The similarity of the rates for both groups is to some extent explained, we feel, by the social and psychological processes which follow the diagnosis. That is, the parents may react toward the younger child in much the same way whether he is severely affected or not.

When employment is obtained it is likely to be of a manual nature. Fig 4. shows that our patients are under-represented in the professional occupations and over-represented in the manual occupations.

A number of young haemophiliacs when interviewed during the survey had felt that leaving school would liberate them from the restrictions imposed by their parents. Such restrictions may in certain patients be justified but are felt to be irksome. Football is very popular in the North East of

England and ~~Table 3~~ Figure 5 shows that 23% of patients were banned, by either parents or others, from playing any sort of football. We hope that this kind of restriction is being lessened now that better treatment facilities are available. They had also felt that leaving school would allow them to leave the stigma of their condition behind them. This is an important matter as the avoidance of stigma requires that knowledge of the person's disorder must be restricted to as few people as possible so that the haemophiliac can pass as normal in as many social situations as possible. We have some evidence that this is what happens in some cases; ~~Table 4~~ Figure 6 shows that 22% of severe patients and 43% of non-severe patients had not told their employers of their disorders. Now, we know that there are many reasons why patients do not tell their employers, some of them entirely justified, and one important reason is that of attempting to be perceived as normal by employers. Many of our patients manage thi process with great skill; an example is a man in his middle twenties, severely affected and with low titre inhibitors, who is on a home therapy regime and has managed to hold down a job without disclosing his condition to anyone at work.

For many, however this avoidance of stigma involves considerable emotional strains. We have seen patients who despite severe walking restrictions, will try to account for their condition without revealing their haemophilia, i.e. car accident or "arthritis". Such people must be constantly on guard lest the true reason for their handicap is revealed. It is within this context that we must work.

What are the implications of these factors for social work with young adults? We have to recognise that improved methods of treatment, especially home therapy and surgery, are making it more and more possible for young haemophiliacs to enjoy a richer social and personal life but this very widening of opportunities brings with it the kinds of problems I have referred

to above and the social worker must be willing and available to give support and help with these problems. Giving help of this kind requires very close working relationships within the Haemophilia Centre. Why should this be so? The first reason is that the nature of the close relationship between physical, psychological and social aspects of haemophilia means that it is not possible to impose rigid professional boundaries on the helping process. Appeals for help with social and emotional problems are as likely to come when physical treatment is being given by the nurse or physiotherapist as when the patient is being interviewed by the social worker. Secondly, the very looseness of the structure as described can mean that the young person faced with a difficult situation may attempt to manipulate one member of staff against another. Effective team work will mean that help can be given by the appropriate member of the care team.

We find that one of our most difficult problems is that of controlling the dependency needs of the young person. We operate a very flexible, almost open-access system and this means that dependency can become a problem. For some people this dependency is enabling; it gives the person something to hold on to during a difficult period and it tends to diminish as the problem is resolved. For others the dependency is disabling in that the relationship with the Centre becomes a substitute for problem solving rather than an aid to such problem solving.

We have found that even in our flexible system there are certain tasks which are more appropriately undertaken by social work members of the team. These tasks are:-

1. Knowledge of and contact with, the whole range of social services, employment services and social security systems which are for the most part external to the hospital. The Centre staff have, in my view, the right to expect that the social worker will be an expert in these areas and will also be prepared to co-ordinate such services for the

individual patient. Our patients have many practical problems which should not be under-estimated.

2. Casework with families with multi-handicaps. The number of such families in each Centre area will not be large. They are, however, extremely difficult to cope with. The social worker would seem to be the most appropriate person to co-ordinate care of such families.

3. An important task of the social worker is to be a resource for other members of the team who may, for reasons referred to above, be dealing with the social and emotional problems of patients. We have a weekly meeting of the nuclear team, as it were, and it is in this setting that members of the team can give advice and support to each other.

We have found that the flexible system I have outlined is necessary to deal with the very complex problems which our patients bring to us. Rigid structures within the Centre can only result in a compartmentalisation of problems which is not, we feel, the best way of helping our patients.

Chairman: Mr Trefor Davies-Isaac is headmaster of Welburn Hall School at Kirkbymoorside in Yorkshire. Within the beauty of an old country house and its estate he and his staff have created a very special residential school for physically handicapped children. He has been of great help to us when faced with a problem not in keeping with our general policy of normal schooling for haemophilic boys.

7 Mr Trefor Davies-Isaac

THE EDUCATION OF THE HAEMOPHILIAC: The role of the residential special school

It has become increasingly common in the last decade to regard the placing of a haemophiliac in a special school as a last resort, and placement in a residential special school as a regrettable decision to be taken only after all else has failed.

While recognising that an increasing number of boys have been successfully accommodated in ordinary schools, it is my task to argue that it is not only desirable but necessary to continue to consider the residential special school as one of the positive alternatives, and that consideration should be undertaken early in the school-life of the haemophiliac, with periodic review. The review should be the responsibility of a team, which should include the paediatrician, the social worker, a teacher, and an educational psychologist but the advice and opinions of other people involved in the total care of the child should be sought and taken into consideration.

Increasingly better methods of identification and diagnosis, and the increasing impact of home-therapy have tended to reduce the effects of haemophilia to the the point where many parents feel able to claim that their son is "virtually normal". It is my contention, however, that some of the other effects which can permanently endanger the development of the educated, adjusted youngster are being ignored. It is unfortunately true that even with the hoped for advent of the age of prophylaxis, there will still remain a small proportion of haemophiliacs whose welfare will depend on placement in a residential special school, just as today one still offers places to diabetic children whose "normal" life is denied them by the inadequacies of their social circumstances.

In considering the residential special school, I should point out that I refer to the school which caters for a wide variety of physical handicaps, including

psychosomatic conditions, and in which the management of the child is governed by clearly defined criteria. It is obviously undesirable to admit a child who is said to be "overprotected"into an environment which is equally overprotective and rigidly structured. Nor would I be prepared to argue my case for the residential special school which caters only for the haemophilic boy, or in which haemophiliacs formed a substantial proportion of the total.

I should like to suggest that a haemophilic boy might be considered for early admission to a residential special school by the following criteria. I would stress "early" in an attempt to redress the balance; far too many of our haemophilic pupils are admitted late, with the consequent compounding of our task. At best, the final result of our work is incomplete, and it occurs to me to ask whether that is the reason why our residential schools are criticised for producing results of a lower standard than one could wish for. I should prefer to see early admission with a consequent early discharge in many cases.

CRITERIA

A child may need a place in a special school:

1. Whose parents are unable to take a sufficiently detailed view of the family predicament to avoid over-protecting and over-compensating the handicapped child.

2. Whose family circumstances combined with severity of the condition would tend to reduce the quality of life, not only of the child, but of the parents and siblings. For example, the difficulty of ensuring, because of work commitment, or other infirmities, that there is always someone at hand to attend to the child's needs.

3. Whose severity of handicap demands such care that the burden of caring for the child presents a very heavy onus of responsibility on the parents, and severely threatens the continuity of the normal relationships of the married couple.

4. Whose family circumstances are poor, and the physical handicap is the decisive factor which tips the scales in favour of residential placement. In many cases, the handicap may be a short-term advantage, securing for the child a better quality of life and better opportunity for learning than he would otherwise have, and which may be denied his "normal" siblings.

5. Whose parents' over-anxious attitude to the handicap would otherwise threaten the continuity of the child's attendance at school.

6. Whose handicap has resulted in a remedial retardation of education and/or maturation, and whose placement in residential school would tend to reduce the level of retardation and immaturity.
Circular 276 (25.6.54) of the Department of Education and Science states: "No handicapped pupil should be sent to a special school who can be satisfactorily educated in an ordinary school. Where a special school is necessary, a day school is preferable if it offers a satisfactory and practicable solution".

This is a fair and considered statement. It is in its misinterpretation that the trouble lies, and in particular in the glossing over the two qualifying clauses. I should like, therefore, to offer, not so much a defence of the boarding special school, but reasons why I believe that many of our present medical and educational colleagues are party to a policy which deprives the handicapped child instead of helping him; a policy which supports the bewildered parent in denying, albeit unwittingly,

the very opportunities which he seeks for his child.

Very many parents, and especially many parents of physically handicapped children would claim that they acted in the best interests of their children, and would even point, with justification, to their own sacrifices on the child's behalf, to their own deprivations and suffering, to the unceasing efforts which they have made so that their child should succeed. It is my task to persuade, as charitably as one can, that some of their actions derive from personal and subjective motives, and are really not to the child's benefit.

Let us agree, for the moment, that there are certainly a few parents who can be defined by common consent, and without exciting controversy, as poor and inadequate parents. Let us ponder for a while on the reasons why good parents resist the admission of their children to the boarding special school.

Parents may oppose residential placement for any of the following reasons:

1. It may simply be more convenient for the child to stay at home, and most authorities will grant parental right of choice. Even if the Local Education Authority disagrees, it is most unlikely to go to law.

2. Parents may often have unrealised guilt complex, and see the total responsibility for the child, with all the attendant difficulties, as their cross in life, which they alone have the right to bear.

3. Parents may wish to preserve the semblance of normality for reasons of status in the community, and believe that this is, at any rate in the short term, best achieved by keeping the child in the normal school at all costs. The costs are not all debited to the parents.

4. Parents may fear, quite naturally, that their child will be distressed by separation. There is no real parallel, incidentally, to be drawn here between the enlightened hospital practice and the residential special school situation. Most of the time, the pupils at the latter are not ill.

5. Parents may fear the adverse criticism of family, friends, and the neighbours, and dread the accusation, spoken or implied, that they are uncaring and rejecting.

6. Parents may fear that they will admit their own inadequacy; most parents, at some time, have quite serious doubts about their effectiveness as parents; the parents of the handicapped child are bound to be haunted to a greater degree.

7. Parents may fear that their children will "grow away from them" - that as older children they will not exhibit the same dependence and indebtedness that children in the close family bond will be expected to display.

It would be interesting to speculate, if time allowed, on the weakness or strength of the wealthier, more privileged classes in this country who have subscribed for centuries to the boarding tradition, and to dwell on the life-work of Kurt Hahn, A.S. Neill, and others but it is not strictly necessary to do so. I should like to propose, instead, that we regard the child with a physical handicap as a child with a difference, and to emphasise that difference insomuch as it creates special needs and special demands. Those demands and needs are less easily met by the normal education system than is often supposed.

There is one further set of criteria which I would offer by which the need for admission to a special school might be measured. These criteria are the needs as I see them, of the child with a degree of physical handicap.

The needs of the physically handicapped child:

1. Close attention to his academic progress, by teachers with the expertise to note and deal with gaps and areas of retardation with particular attention to the basic subjects, and maths most of all - the first casualty of any absences. It is important to measure performance regularly and to compare with ability. It should always be possible to withdraw a child for individual attention at any time that the need is indicated.

2. Expert guidance and encouragement in physical education, games and movement activities. This is almost taken for granted in special education today, although my predecessor was thought to be eccentric when in 1962 he sought to add a specialist Physical Education teacher to the establishment. With the strong support of doctors and physiotherapists, he got his P.E. teacher.

It is nearly always possible to give the handicapped child his first meaningful experience of success in the very area which had previously been outside his experience, or in which he had known frustration and failure. This is true whatever the physical handicap. It is not only rehabilitating, and complements the work of the physiotherapist, but is of positive value in building up morale and self-image.

3. Good support for progress towards whatever degree of independence is possible, depending on the degree of handicap. This training for independence needs to include self-care, hygiene, social acceptability, a positive outlook on the youngster's role in the community, with a sound sense of his responsibilities, as well as a knowledge of high rights as a citizen. He needs to be encouraged to develop a vigorous, even aggressive determination to succeed in life, and to be persuaded that it is not good enough to be as good as the normal contemporary : he must be better.

4. He deserves the opportunity to grow up with his peers. Quite often the supportive role is played by the parents when the physically handicapped child attends a normal school, or even day-special. The child is so often the focus of attention, and is the solitary youngster with a phalanx of adults in attendance. Parents often go to great lengths to give their child the benefit of for example trips to the swimming baths, shopping or youth clubs. What the youngster is often missing is the educating influence of his contemporaries. I mean this in the broadest sense, for with the good he will also learn the undesirable things- but he will be given the opportunity, which is the right of every child, to develop a balance morality, and learn what is fair and honest. I am convinced that, however hard and long my colleagues and I work as school, the children themselves are the strongest force for developing each other's characters.

5. The child needs to be thrown on his own resources to a greater extent than is possible in the normal caring family. This sounds callous, and indeed one feels callous in operating this policy, but it pays devidends in the development of self-reliance, and learning how time can be usefully used. It is a policy which we described

as"benevolent neglect", and is not to be confused with the old story of "here's a shilling to go to the pictures", or sending the children to Sunday school to ensure a peaceful Sunday afternoon. It may be hard on the children at first, but life is hard, and harder still with a handicap to contend with. We want our children to grow up with a will of iron and the emotional hide of a rhinoceros, able to meet any adversity with resolution and determination.

6. He needs to be segregated for some time, and to a diminishing extent, in order to be able to come to terms with his own handicap. His life in the special school will be one of no concessions and unremitting demands on his abilities. I know of very few parents who could display that objective attitude towards the child and his difficulties which is necessary to develop the necessary physical and mental attitudes. I know of no normal school where concessions are not constantly made by staff and other pupils to the detriment of the handicapped child's development.

"..... the present of these physically handicapped children in the school made no difference to the attitude of the other children except to emphasise the consideration and kindness they showed their handicapped fellows <u>for whom they did all they could</u> ....."

"..... non-handicapped children go out of their way to help their unfortunate class-fellows".

(Health of the School Child Department of Education and Science).

If we are to achieve the acknowledged aims of enabling the haemophilic boy to come to terms with his handicap, to receive education suited to his ability, and to develop into an adjusted balanced adult, as independent as possible, I contend that we need

to consider very carefully before rejecting as no longer necessary the benefits which can accrue from attendance at a residential special school.

SUMMARY

Experience with physically handicapped children including haemophiliacs suggests that among their many and varied needs the following are of special importance:

1. close attention to academic progress by teachers trained in the early recognition of areas of retardation especially in basic subjects.

2. expert guidance and encouragement in physical education and games, complementing the work of the physiotherapist.

3. training for independence and fostering of a determination to succeed.

4. the opportunity for children to grow up with, compete with, and be guided by their peers.

5. recognition that children must occasionally be thrown on their own resources in order to develop self reliance.

6. realisation that segregation from caring parents and protective teachers too ready to grant concessions will help, rather than hinder, children learning to come to terms with their handicap.

NEWCASTLE IXa (LINK)

Chairman: Dr Desmond Dunleavy is Senior Lecturer in Psychiatry at Newcastle University, joining the haemophilia team in 1975. Apart from his help with specific problems of management, his common sense approach and fresh thinking have enabled us to examine some of the problems facing severely affected haemophiliacs from new angles — whether he has been of more assistance to staff or haemophiliacs is debatable*

* Footnote: Dr Dunleavy was unable to be present on the day of the meeting as GRO-A

GRO-A

8. Dr Desmond Dunleavy

THE PSYCHIATRIST IN A HAEMOPHILIA TREATMENT CENTRE

People are usually anxious when they are confronted by a Psychiatrist as they think that somebody, if not they themselves, must feel that they have some form of mental illness or are at the very least a little unstable. If this is true what can a Psychiatrist possibly have to offer in a Haemophilia Centre? Is there something about Haemophilia which makes sufferers more liable to psychiatric illness which makes his presence obligatory? Immediately, therefore, let me state that it is my opinion that Haemophiliacs are no less or more likely to develop psychiatric illness than the rest of the population. A number of Haemophiliacs will develop some problems related to living as has already been mentioned by other speakers, but this should not be classified as mental illness. However, a case can be made for the attachment of a Psychiatrist to such a Unit, and I will base it on my experience in the Newcastle ~~Unit~~ Centre over the past few months. I will briefly discuss two areas, firstly, the case for involvement and then problems which I found loom large in such a ~~Unit~~ Centre, i.e. those of dependency and drug abuse.

CASE FOR INVOLVEMENT

One of the first things that a Psychiatrist does when confronted by a patient and his problems is to take a lengthy history in order to see which factors in the patient's background may be important in producing his present symptoms, and his findings will influence his plan of management. Assessment has therefore to take account of many interacting variables but it is vitally important to know the norms for the particular group with whom one is involved. Involvement in the Haemophilia ~~Unit~~ Centre inevitably leads to a greater knowledge of the illness itself, and to an awareness of the major adjustments called for by the patient in his work and ordinary social life.

With experience it is possible to separate off problems due to the illness itself and problems which arise from the patient's own personality. There is some interplay between these factors but one's understanding of the total problem is that much better from seeing a number of people who suffer from the same illness. As I have become aware of the background problems of Haemophiliacs in recent months, I find it easier to assess whether their problems are arising as the result of mental illness or just the individual's attempt at adjusting to his illness.

PROBLEMS CREATED BY SUCH A UNIT

In the not too recent past Haemophilia was a crippling, not to say frequently lethal illness, and if you suffered from it one was not expected to lead a productive or active life. Modern treatment methods towards which the Haemophilia Treatment Centre is geared, help the individual sufferer to take a more active part in society. To some extent he is put in a difficult position because although he has an illness, he is no longer able to take a "sick role". He is at one and the same time encouraged to be independent, and yet independent in a very real sense on the immediate availability of treatment for his continuing survival. Some individuals have problems overcoming their dependency needs and will inevitably become more dependent on the ~~Unit~~ Centre, and as a result make more demands on its services. The ~~Unit~~ Centre itself is able to tolerate these demands, but unfamiliar medical staff in accident and emergency rooms may think the patient over demanding or even simulating pain and an escalating situation develops when the doctor becomes angry and the patient in turn becomes angry and upset and even more demanding - result:chaos. There may be a tendency also at times of stress to fall back on complaints of increased pain with demands for analgesics or some other form of help or support. Dependency can be expressed in other ways: in apparent unwillingness to obtain employment and a passive life style. This sort of problem can only be managed over a long time period and requires a great deal of tolerance and sympathetic understanding; walking a tightrope

between firmness on the one hand and total acceptance on the other.

DRUG ABUSE

One other problem which I have become aware of is the tendency for some individuals to increase their intake of analgesic or pain killing drugs which they use for psychological purposes rather than their direct therapeutic effect. The amount of analgesic which some people ingest over a short time period can be quite dramatic with large escalation in dosage from week to week. This then causes distress to the patient himself, his relatives and his medical attendants (who fear addiction) and consequent deterioration in the patient's personality. The problem mainly arises, of course, in the adolescent haemophiliac and is part of the wider problem of adolescent drug abuse in the general population, rather than something which is specific to haemophilia. Availability of drugs is thought to be one of the factors responsible for continuation of adolescent drug problems and the young adult haemophiliac has ready availability to potentially habit forming drugs. The adolescent drug abuser is quite different to the drug addict, and there are many reasons why this problem arises. It can be part of the normal adolescent rebellion against parents or authoritarian figures; part of the seeking for an identity; identification with a special group; or the assertion of one's own independence. The problem therefore must be looked at firmly within the context of adolescent maturation. In the case of the haemophiliac some extra problems may be imposed on the maturing young adult who now has to come to terms with his illness in relation to job opportunities and his role as a maturing sexual adult. The problem can only be dealt with therefore on an individual basis. Fortunately it is usually a transient phase in development, and the Psychiatrist's role is one of damping down anxieties in the individual himself, and his relatives. Exploration of the situation and simple explanations will relieve the immediate problem but reducing the drug abuse itself is a long laborious problem which will need the active collaboration of many different agencies. There are a

number of pointers towards the person who is more likely to develop problems in this area but attempting to build a stereo-type can be dangerous. Addictive problems in the Haemophilia ~~Clinic~~ Centre are not a major problem at the present time but further research and experience will be needed before a definite statement can be made.

SUMMARY

I see the Psychiatrist's function in a Haemophilia ~~Unit~~ Centre as being peripheral. He is there to offer an opinion in consultation when necessary; to take part with others in the management of a small number of patients where his help may be appropriate; but he will not play a significant part in the day to day running of the ~~Unit~~ Centre for the great majority of patients. Most of the problems he deals with will not be related to Haemophilia itself, but only come to light incidentally because the patient is attending the Centre for the treatment of his primary disorder.

Chairman: Mr Rennie Porteous, a Senior Lecturer in the Department of Child Dental Health, was a founder member of the team in 1966. As a result of his work in general dentistry and conservation, the Newcastle children with haemophilia can probably claim the finest occlusions in the country.

9 J Rennie Porteous

DENTAL CARE FOR HAEMOPHILIC PATIENTS IN NEWCASTLE

One of the major recurrent threats to the haemophilic patient throughout his life-span is from dental problems.

Dental care of the haemophiliac should begin as soon as possible after diagnosis and should be prevention orientated. In Newcastle all patients attending the haemophilia clinic are routinely examined by the dental surgeon on duty. In this way we check on the patient's progress and if problems present themselves they can be resolved in joint consultation. The maintenance of routine dental treatment either in my own department or by general dental practitioners is also ensured.

I would like to discuss management and treatment of haemophiliacs in the following order:-

1. Prevention
2. Conservation
3. Endodontics
4. Oral Surgery

1. PREVENTIVE CARE must include:

(i) Dietary advice:-

The one certain way of reducing dental caries is to restrict the intake of readily fermentable carbohydratesto meal times. Parents must be warned of the dangers of comforters and comforter bottles and secondly the dangers of indiscriminated "between meals snacking".

Figures 1,2,3,

(ii) Oral hygiene instruction:-

It is doubtful whether toothbrushing alone will prevent dental caries, but there is no doubt that toothbrushing prevents periodontal disease. In this way the haemophiliac can reduce the dangers of tooth loss from gum disease.

Figures 4,5,6.

(iii) Fluorides:-

Where there is less than 1 part per million (ppm) fluoride in the drinking water we are prescribing fluoride tablets to raise the level ingested to 1 ppm/day in patients aged 0-12 years. Fluoride incorporated into developing teeth will reduce the caries incidence by as much as 40 - 60%

In the older patient, rather than prescribing fluoride tablets the topical application of 2% sodium fluoride or acidulated phosphate fluoride is instituted in an attempt to reduce further the dental caries problem. We are now starting a home-treatment routine where those patients willing and able to co-operate rinse 2 mins daily with 8 mls of 0.05% solution of sodium fluoride. Results so far are encouraging and suggest as much as a 30% reduction in caries.

We advise all patients to use fluoride toothpastes as part of their oral hygiene.

(iv) Orthodontic treatment:-

The development of the occlusion is carefully watched in an attempt to minimise overcrowding problems particularly in the third molar region. Each case must be assessed separately and extractions are contemplated only where the physician concerned considers such treatment is possible. Tooth movements using

appliance therapy can be undertaken using removable or fixed appliances. In this way irregular alignment of the teeth can be avoided, thereby reducing the risk of periodontal disease.

Figures 7,8,9,10,11,12.

2. ROUTINE CONSERVATIVE TREATMENT: may represent a great problem to the haemophiliac. Deciduous teeth, on the whole, are not very sensitive and can usually be restored without too much difficulty. It is in the permanent dentition that pain during cavity preparation becomes an important factor. In the normal patient local anaesthetic is usually sufficient to control this pain. The administration of a local anaesthetic to a haemophiliac is an extremely hazardous procedure and at least two deaths following inferior dental block anaesthetics have been recorded (i.e. Archer & Zubrow (1954) and Parnell (1964).

In the past in Newcastle we have resisted the use of infiltration anaesthesia and banned the use of regional local anaesthesia. Now with improved replacement therapy we are using local infiltration anaesthesia for out-patients attending the Dental Hospital.

Figures 13,14.

In cases where there is extensive caries, low pain threshold and poor co-operation, I still prefer to admit the patients to hospital and carry out treatment under a general anaesthetic. Naturally one recognises the hazards of passing an oro-pharyngeal tube but up to the present time we have had no problems. In addition, all the treatment can be completed in one session and usually the only prophylactic treatment prescribed is E.A.C.A. sometimes following one injection of a factor VIII preparation.

3. ENDODONTICS: may be carried out for the haemophilic patient. In this way dental abscesses can be treated both in the deciduous and permanent dentitions without extraction.

Figures 15,16.

4. ORAL SURGERY: for the haemophilic is well documented. In general, dental extractions are better planned electively when all the supporting services are available.

In Newcastle we have the following routine:

(i) impressions are recorded on admission for the construction (only if necessary for instance in antibody patients) of metal cap splints to carry supporting flanges to enable sockets to be protected if bleeding persists. Splints are not used routinely, and are very rarely required.

(ii) replacement therapy as decided by the physicians, usually one injection of factor VIII preparation and an antifibrinolytic cover, after excluding antibodies.

(iii) extraction of the teeth with minimal trauma.

(iv) the sockets are packed with gel-sponge soaked in thrombin and sutures inserted to consolidate sockets and encourage the development of a stable clot. The patient is discharged the following day on an antifibrinolytic drug and an antibiotic.

In conclusion I would like to stress the need for a preventive approach to dentistry for the haemophilic patient, such as we are seeking to develop in Newcastle.

COMMENTARY FOR ILLUSTRATIONS

1. Child sucking comforter or "dummy"

2. Caries present in this 18 month old child's mouth due to dipping the comforter in a syrup.

3. Results of Vipeholm dental caries study (Gustafson et al 1954) Individuals receiving readily fermentable carbohydrates with meals (represented by dotted lines) have low caries increment. However those having unlimited between-meals snacks (represented by continuous lines) have a high caries increment.

4. Patient with poor oral hygiene, gingival inflammation. Claimed to brush teeth regularly.

5. Teeth treated with disclosing solution indicated oral hygiene procedures were limited to the mesial aspects of 1/1. Crowding in the dentition could be an additional difficulty in oral hygiene procedures.

6. 2 years later following extraction of first premolar teeth to relieve crowding the occlusion is developing soundly. Oral hygiene instruction and technique good with resultant healthy gingivae.

7. Patient with crowding, lateral incisors inside the upper arch. (mirror view)

8. Same patient from the side with the teeth in occlusion.

9. Radiograph confirming present of all teeth including 3rd molars.

10. After treatment involving the extraction of all four first permanent premolars and proclination of the upper lateral incisors.

11. Radiograph confirming development of the occlusion.

12. An extreme example of third and fourth mandibular molar impactions (fourth molars are v.rare)

13. Haematoma developing after an infiltration local anaesthetic for the conservation of an upper molar.

14. Complication here was trauma, due to closure of the teeth to buccal mucous membrane resulting in a potential pathway of infection to the main haematoma.

15. Root canal therapy completed for a patient with inhibitors.

16. Continuing improvement of periapical state in the same patient.

17. Patient after total clearance, note healthy sockets and the presence of resorbable sutures.

COMMENTARY FOR ILLUSTRATIONS

1. Management and treatment of haemophiliac patients.

2. Preventive care.

3. Child sucking comforter or "dummy".

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(Courtesy of P. Bradnum F.D.S.).

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Chairman: Mr Brian Fleming is a Consultant Surgeon in the Royal Victoria Infirmary and recently appointed Clinical Sub Dean in the University of Newcastle upon Tyne. He has worked with haemophiliacs for many years and is responsible for the post-graduate teaching of surgery and the haemostatic disorders.

THE ROLE OF THE SURGEON

I am very pleased to have the opportunity of speaking as a member of the team responsible in Newcastle for the total management of the haemophilic patient, and I speak as a general surgeon, not as all the other things that Peter mentioned. I think it is important that there should be a general surgeon as a permanent member of the team because of course haemophilia does not protect the individual against those ills and conditions which are amenable to surgical correction, such as hernias, varicose veins, gallstones and duodenal ulcers. It is also important that the surgeon should be one man, one individual who takes an interest in ALL problems of bleeding in surgical patients and thus is able to accumulate an experience not only of the bleeding disorders but also of individual patients and of working with other members of the team.

Now the role of the general surgeon is one which has changed in recent years. Formerly we were called in two particular circumstances. In one we were called to deal with the complications of haemophilia itself, usually in the form of haematomata occurring in the soft tissues. These haematomata were very unpleasant as they grew slowly in size, compressing and destroying normal tissues. The method of treatment was to lay them open widely, to remove the clot, to secure those bleeding points which could be secured and to embark upon a long train of packing with various antiseptic solutions. At best we finished up with fibrosis, at worst with cripping deformities. The other set of circumstances in which the services of the general surgeon were sought were for life-threatening complications of haemophilia, notably of course bleeding from duodenal ulcers, and indeed those of us who were involved in this kind of surgery were regarded somewhat askance by our colleagues who regarded our efforts as - as it were - flying in the face of fortune, often taking what appeared

to be unnecessary risks. I like to think these days are past and gone. They have passed because of the advent of cryoprecipitate and the freeze-dried Factor VIII preparations, so much so that we are no longer confined to offering surgery only for complications of haemophilia or for life threatening conditions. I rely on Peter Jones and the rest of the team to provide for me the patient whose haemophilia has been temporarily suspended in the immediate pre-operative phase, during operation and for a sufficient length of time post-operatively to avoid the complications previously encountered and my remarks of course apply not only to haemophilia itself, but to other bleeding disorders such as von Willebrand's disease. In fact I make a contribution here by feed-back of information, because I know of a number of patients whose behaviour on the operating table is that of a bleeding diathesis although no specific factor abnormality can be measured in the laboratory. These patients require the precautions which we apply to the haemophiliacs, to the Christmas disease and von Willebrand's patients and here again I refer back to Mr Oxley's comment this morning. These are the occasions where the practicality of the situation over-rules the laboratory findings.

We must never, however, achieve complacency in these situations. We must never rely solely on the Factor VIII preparations to do our job for us in surgery. We must always be meticulous in our technical haemostasis. Professor Ingram this morning referred to the most noble haemophilia carrier, Queen Victoria. I should like to do so again this afternoon to illustrate my point. About 100 years ago France was rules by the Emperor Louis Napoleon and his consort the Empress Eugenie. They were not popular with the Paris bourgeoisie and not popular with the press who lost no opportunity to deride the Royal couple. It was said that, during the state visit of Queen Victoria to France and the obligatory performance of the two national anthems at the Opera, the parvenu Empress Eugenie looked round for her chair and sat down, whereas the English Queen merely sat down because she knew

there would be a chair there. Most surgeons in their attitude to bleeding tend to be Queen Victorias; they know that there is a chair there, they know that bleeding will stop. But in dealing with the haemophiliacs we should emulate the Empress Eugenie and ALWAYS check that ~~there was~~ a chair is there.

One minor role played these days by general surgeons in haemophilia is concerned with genetics. I am more frequently being requested to carry out the operation of vasectomy in male haemophiliacs who wish to protect their grandchildren, in other words to avoid producing haemophilic grandsons.

What of the future? Professor Macfarlane this morning pointed out the natural expectancy of life in the haemophiliac is approximating that of the non-haemophiliac and therefore, inevitably, haemophiliacs as they get older will undergo the wastings of age common to us all, namely enlargement of the prostate gland and, I fear, an incidence of malignant disease. In the future I look to a time when haemophilia is merely as incidental as is diabetes today. We will then be able to offer any form of surgery to haemophiliacs as we now do to the non-haemophiliac and with the same degree of safety.

Chairman: Mr David Stainsby, who came to Newcastle from Oswestry and Sheffield, is a Consultant Orthopaedic Surgeon. He has been a member of the haemophilia team since 1968 when we met over a pseudotumour one Saturday afternoon. He plays a major role in advising the Centre staff about the management of the many and varied acute and recurrent lesions of haemophilia, and is responsible for the orthopaedic surgery programme.

11. David Stainsby

THE ROLE OF THE ORTHOPAEDIC SURGEON

During the last eight years in Newcastle, 21 orthopaedic operative procedures have been carried out on haemophiliacs. Four were routine operations unconnected with the haemophilic condition. Fifteen were for recurrent bleeding into joints and a painful haemophilic arthropathy and two procedures were done to remove enlarging pseudo tumours.

The routine operations were: laminectomy, meniscectomy, a Keller's procedure for a painful hallux rigidus, and an elongation of a tendo achilles in a boy born with a club foot deformity. Adequate factor VIII cover was given and operation and subsequent progress were uneventful in all four cases.

Of the fifteen procedures done for haemophilic arthropathy nine were synovectomies, two of the elbow and seven for the knee. Three knees and one ankle have been arthrodesed; one patient has had a patellectomy, and one total hip replacement arthroplasty has been carried out.

Both patients who had synovectomies of the elbow joints had advanced joint disease. They had persistent pain and frequent haemarthroses and their elbows were swollen and movement reduced. Figure shows an x-ray of one of the elbows showing the distortion of the joint outlines. At operation the thickened synovium was found bulging through the capsule and when the joint was opened the brown stained thickened synovium was seen. After removal of the synovium the erosions at the articular margins and the fibrillation and staining of the joint surfaces became apparent. Both patients have benefited from operation. Pain has been relieved, haemorrhage is now infrequent and elbow movement only slightly reduced from the pre-operative state.

Seven knee synovectomies have been carried out on six patients who had an age range from 12 to 25 years. All these patients were disabled because of repeated bleeding into the knees uncontrolled by conservative measures. Before operation they all had at least 90% of movement and no angulatory deformity was present. All had x-ray evidence of arthropathy, but without exception the extent of the joint damage seen at operation was much greater than that suggested by the x-ray. The x-ray of this 25 year old man's left knee (Figure) shows a little squaring off of the medial, femoral condyle and marginal lipping and erosion. It looks a reasonable joint. At operation gross synovial staining and thickening were found. After synovectomy, the lateral femoral condyle looked reasonable although the articular cartilage was stained, but the medial femoral condyle showed marked fibrillation of the articular cartilage and central depression due to damage to the underlying bone.

Two patients had gross x-ray changes and the patella had produced a groove between the femoral condyles. However, as they had over 90% of movement synovectomy was carried out rather than stiffening the joint. Complications occurred in three of these patients on which we carried out synovectomy. One had a stiff knee two months after operation and manipulation of the knee was carried out under anaesthesia. This was a mistake as a supra condylar fracture of the femur resulted. The fracture subsequently united but the knee has only regained 40° of flexion. One patient bled into the joint cavity following synovectomy and a further operation was required to remove approximately a litre of clot from the knee. Subsequent progress was uneventful and 90° of flexion movement has been regained, giving a satisfactory result. Another knee did not mobilise well after synovectomy regaining only 50° of movement.

Now, despite the fact that these synovectomies were carried out on knees

with considerable damage to the articular cartilage the results have been remarkably good. All seven patients have had relief from recurrent haemorrhage and pain and only two have lost significant movement.

Arthrodesis. For three seriously destroyed knees (Figure) and one ankle, arthrodesis has been performed. In all cases, compression arthrodesis has been carried out. Figure shows the clamp and probe in position; all four arthrodesis united without complication. Figures show the ankle and the x-ray of it soundly fused.

One patient aged 20 years had pain in the knee which seemed to come from the patello-femoral compartment. At operation a central erosion was found in the articular surface of the patella and so it was removed. The pain was relieved following this procedure and knee mobility regained, but occasionally bleeding into the joints still occurs.

Arthroplasty (that is the insertion of an artificial knee joint) has not been attempted. The hinged arthroplasty is considered to be unsuitable and as yet in Newcastle we have insufficient experience with the duo condylar type of prosthesis to justify its insertion into a haemophiliac. The Charnley hip arthroplasty is however now regarded as a standard and reliable orthopaedic procedure. We have carried out one hip arthroplasty. This was done for a 52 year old man with a very painful stiff left hip. The x-ray (Figure) shows the marked distortion of the femoral head. Arthroplasty was carried out a year ago and so far the result is very satisfactory with relief of pain and restoration of movement.

We have had to deal with two pseudo-tumours. The first was a 26 year old man who had had recurrent bleeds into his calf muscles. A further bleed occurred into the right calf in December 1969, but despite immobilisation and factor VIII the bleeding continued and the overlying skin necrosed. Figure shows the state of affairs when he was

transferred to the Newcastle Centre in January 1970. The figure shows the full length of his calf. With factor VIII cover this pseudo-tumour was explored and the clot removed. Following suture of the skin flaps subsequent healing was uneventful.

The second patient was aged 21 and had had meniscus (cartilage) removed at another hospital without it being realised that he had any clotting abnormality. Post-operatively he bled into the joint and from the operation wound, and it was then found that he was a mild haemophiliac. Bleeding was eventually controlled but he was left with a persistent painful swelling beneath the operation wound on the inner side of the knee. Five months after his meniscectomy this swelling was explored and found to be a pseudo-cyst. Figure shows the cyst measuring about 4 cm in diameter overlying the medial joint line. When cut into it contained a blood clot and appeared to be attached, and to be arising from a small cortical defect at the margin of the tibial condyle indicated by the probe. Presumably this was caused by slight damage to the bone surface at the time of the patient's meniscectomy. The cyst was removed and post operative progress was uneventful.

It will be seen that our surgical policy has been a rather conservative one. Particularly as regards synovectomy and we have been operating on joints severely damaged by repeated bleeds which occurred before adequate factor VIII replacement was available. Now that there is an active home therapy programme in progress we are hoping that joint damage will be prevented by prompt control of any bleeding episode and then this type of salvage surgery may rarely be required.

DISCUSSION

Q. I would like to know the length of time that you have a splint on for, if you were using a back splint or anything like that?

Miss Cole: We tend to use it until the swelling and pain have gone down or we can use another splint like the Los Angeles splint with the other bit on the front, to help strengthen weakened muscles. The fact that this splint is not bandaged on from top to bottom means you have got to use the thigh muscles, and we would leave it on until the quadriceps have reached a par 4 which means they do not need assistance.

Q: Mrs Lovie said during her talk that she gave support to the parents and young haemophiliacs during the formative years. How much attention does she given to the talks at the local haemophilic group.

Mrs Lovie: As you know, I have only recently become involved with haemophilia. I have been to the meetings and it is a case of me learning from them. This is the way it works.

Chairman: I think that Susan Lewis, who went out on the Survey, can perhaps answer this specific question.

Mrs Lewis: Yes. I think at the time we did our Survey, which was started in 1973, the local Haemophilia Society Group was not very active and this was partly due to the geographical distribution of patients. Obviously when you have patients 200 miles apart it is very difficult. Also with such a distribution it is very difficult for everyone to be aware that there is a Group. When I performed the Survey I did make a point of asking the question of how many people were members of the Society. For everyone who was not a

member I sent the name and address (with their permission) to the local branch, so that hopefully we will see the return of this in the next year or so. We already participate in Society discussions at their invitation.

Q: Dr Jones, I was delighted to see the number of people you have on home treatment. One of the things that concerns me (as the secretary of our own group in Cambridge) is the very high incidence of jaundice you seem to have picked up; can you account for this please?

Chairman: The questionner has just pointed out that an outbreak of hepatitis has been seen in the United Kingdom following the introduction of commercial Factor VIII concentrates on which some Centres are unfortunately almost totally reliant (apart from the use of cryoprecipitate for inpatient therapy), and up to about eighteen months ago we did see hepatitis. In Newcastle we saw 22 cases of hepatitis, a small proportion of which became Australia antigen positive. I think that this can be explained for two reasons. First, these products were really coming into virgin soil. The blood of a different population was being used for the first time in the United Kingdom to treat haemophilic patients. The second reason was that at the time that the first batches of these products were introduced into the country the sensitivity of the testing methods for the hepatitis associated surface antigen were not as sophisticated as they are now using Aus RIA II (Australia radioimmunoassay). We have not seen an episode of hepatitis in the past, I think, nine months to a year, except for one who appeared last week. In this case, because of the relative incubation period, I think it is more likely to be linked to cryoprecipitate than to our use of commercial concentrates. Hopefully therefore the disease which

we saw and which, although disturbing did not bring any of our patients into severe detriment, has passed. Of course we will see sporadic cases, but yes it was very disturbing. Hopefully it is over but it is worth remembering that some people would say that it is better to get a mild case of hepatitis and therefore become immune in the future.

Q: I would like to make a general comment on some of the things I have heard here today and I think that all the people in this audience has been struck by this whole performance (if I may use that word) that you have presented. This collaborative effort, we have been very impressed by it and I would like to ask you a few general questions about it. First of all how has this effort been received by the other hospitals around in your region? Have you managed to get around the rivalry one so often sees within the medical community. Secondly what do you think about the leadership of such an institute as you now have up in Newcastle; who is the right person to select? Should it be a doctor or could it be one of these other members that seem to co-operate so well with you in this project.

Chairman: Thank you very much Dr Evanson. The Norwegians are renowned for their frankness! I am not sure I can fully answer all of your questions. The first one was that Dr Evanson wondered what the reception was amongst the other hospitals in the Newcastle region. I am happy to say that we have the greatest co-operation and friendliness with all our colleagues in the regional hospitals. This perhaps is surprising but I think that when people see what haemophilia can mean to families, and they see it first hand and some new methods come along to treat it (for instance cryoprecipitate or the Factor VIII

concentrates) then they are only too ready to help. The way we run things in the region is to work with our Haemophilia Centres and associated Centres each of which is staffed by a pathologist, physician, paediatrician and a dental surgeon and they take care of the haemophiliacs within their own immediate area. Newcastle very strictly and with total agreement runs the home therapy and the major surgery programmes. So we work happily and hopefully will go on working happily together.

The only comment I would make about your second remark about leadership is that I think that the extent to which haemophilia has become involved with every single aspect of living means whoever is going to direct a Haemophilia Centre must be full-time (nor nearly full-time) in order to do it. I really will not comment further except to say that I could not work without the people you have seen speaking this afternoon, but I think that you must have somebody in general administrative charge.

Dr Katherine Dormandy

Just a quick question. I would like to know to what extent the patients, particularly the adult patients, collaborate with the dental regimes. You see we have a lot of adult patients who just have to be chased and chased and chased and chased. And so it does not matter about the willingness of the Dental Department or the efforts that the Sisters and the secretaries put into it, they still do not keep their appointments. Does that happen in Newcastle?

Mr Porteous: We have exactly the same problem in Newcastle particularly with our adults. I think that there is a feeling that one begins with children and builds from there. Hopefully we are now beginning to see a change in attitude nationally within the population about dentistry and I think that this will grow. But until such time as these children that we have now become adults we are going to be left with the problem of years gone by that the adults <u>are</u> difficult. With someone prepared to take an interest in them their interest is increasing.

Dr Rosemary Biggs: I should just like to make a small comment about what Dr Evenson said about who should lead such a team. I think the thing about a leader is that they choose themselves. It is a total myth which is current in our computerised Western society that you can have a central edict which says let there be so many Haemophilia Centres and let them be directed by, lets say, a physician. When you do this you say to some doctor "Will you have a Haemophilia Centre" and he says "Yes" and then he goes away and does what he is doing anyway; he certainly does not have what is meant by the comment of a Haemophilia Centre. So that the people who become leaders of Haemophilia Centres are people who have an interest in this <u>certain field</u>. It could be a dentist, it could be a paediatrician, it could be a pathologist, it could be an orthopaedic surgeon. It is entirely a question of the personalities of the people concerned and I think this evolutionary attitude is extremely important because you could say "let there always be a Haemophilia Centre in some particular town". Well that doctor might move, and there is absolutely no guarantee that the doctor who comes to replace him will have the slightest interest in the haemostatic diseases whatever. Three factors need to be taken into account by any sensible authority in re-planning the re-allocation of

of Centres, and with the administration in this Country we are developing a system by which the number of Centres is listed, and is going to be listed every year allowing for changes in evolutionary development which occur in the society as it progresses. But I think this is absolutely not a question which can be dealt with by a central planning authority.

Dr Davies — Can I first of all say how interesting I have found the afternoon and I think it is a great tribute to the Newcastle team whom we have heard led by Peter Jones. I have always had a Bete Noir about haemophilia and the education and the upbringing of haemophilic children and I have never believed that they should be segregated from the normal child. I think that the haemophiliac is peculiar in that his handicap is intermittent to the outsider, and that many of the children who go to special schools have a permanent and visible handicap. This sets them aside in some way from the haemophilic child and I think that nowadays with the increasingly mobility and increasing ability to (using the horrible American word) "normalise" the haemophilic boy, it is even more important that he is integrated totally into society from an early age.

Mr. Davies-Isaac — Briefly, this is true as a generalisation but it is true not only of children with haemophilia but children with other physical defects as well. It is also true that there are children with physical handicaps who can only benefit by a special school. It is also true, I think as you have seen this afternoon from other speakers, that there are haemophiliac children who have suffered educationally through not having been to a special school. Whereas you may say they have achieved

"normality" in one aspect by remaining within the normal range of education, have they achieved "normality" at the expense of missing out so badly on their educational opportunities during the time that they were of school age? Which is more important? A bad special school is simply a bad school, there is no such think as special schools which are all bad because they are special.

Q. I take the point about special schools but surely the improvement in treatment that the children are receiving now is going to mean that they lose less schooling time than those who are now adult?

Chairman: I think that is absolutely true, but I think that when you are dealing with a number of families with haemophilic children (and dealing with the whole family) there will occasionally be a time that you will have, for the reasons that Trevor Davies-Issac discussed in his talk, to think seriously about taking one or two percent of your population and asking people like him to care for them in a special school rather than a normal school. But yes, I think that we are totally in agreement that the majority of haemophilic children should go to normal schools.

Q. I wonder if you could tell me is there an actual schedule for using cryoprecipitate or concentrates for hospitals?. Is there a schedule telling them the exact way to give it and the correct times and things like this, or is it just done by the whim of the hospital?

Chairman: Yes, very much so. The first schedule I would suggest is in Dr. Rosemary Biggs' book which includes the full dosage regimes for all the factor VIII and other products which are used in the treatment of patients. In our particular region we have a "haemophilia handbook" which all the hospitals dealing with haemophilic patients have at their disposal. Hopefully the junior staff, in spite of having so many other things to do, ~~and all of their overtime claim forms to fill in now a days,~~ find time to read a bit of it and give the right dose to patients.

Q. Is there any way of asking them to read this book? In some sort of hospitals they do not seem to have heard of the methods.

Chairman: I am not sure of the price of your recent book, Dr Biggs, but I suggest that a donation for the new edition might help. Would you like to comment on that?

Dr Biggs: Well I think it is a very difficult question. I do not think there is any way of making people who are far too busy with other things and only see one haemophilic patient every three months ready about it. Nor is it possible to make a situation where the treatment at that hospital is going to be 100% safe, because the house officers will rotate as fast as the haemophilic patients come in. I think that the only way that this can hopefully be developed is by a new method of co-operation, if it can be evolved between the parents and the patients and their medical staff. Maybe the parents of patients who live in a place like this ought to go and take a sort of course with Dr Jones' book (if I can retaliate!) and be 'graduates' in the care of haemophiliacs. They should then go to these hospitals and instruct the medical

(9)

staff how to treat their children, and be quite determined about this. Quite a lot of the parents of our patients are perfectly capable of doing this! Maybe there ought to be holiday courses, say at some of the special school like Lord Mayor Treloar College, where people can go and get instruction on how to know exactly how often the patients should be treated; these hospitals should develop some of the things like the Canadian people do, all the patients know where the cryoprecipitate is and know exactly how it should be used. They are instructed to go to the hospital and get this out and then simply deliver themselves and the stuff to the casualty department and say "This is what I am to have". It is organisations of this sort which will have to be developed.

Q. Please could you comment on an aspect of the home therapy programme? Mr Oxley mentioned a patient on home therapy who had antibodies.

Chairman: It is correct that we do not with-hold home therapy simply because of antibodies. However very much depends on the level of antibodies and their response to treatment. One important thing is the clinical response to treatment. If bleeding always stops when factor VIII is given very early in the course of a bleed, then naturally the quality of the patients life is improved. All one can say is that, for some antibody patients, routine early treatment of bleeds with factor VIII works. It works clinically and it stops early bleeds and that is of course the basis for home therapy.

End

Closing remarks : Dr Jones

In thanking you for your courtesy and discussion and in wishing you an enjoyable conference I would like to end by quoting the words of a man deeply involved in another minority group. In spite of the recent advances in care some haemophiliacs must inevitably feel lost in the rarity and unending nature of their disorder. Dr Martin Luther King, speaking in Los Angeles, recalled the words of an old Negro slave which I think are apt in the present state of the haemophilia game . . .

> "We ain't what we ought to be and we ain't what we want to be. But thank God we ain't what we was."

Acknowledgements

The speakers wish to thank Miss Margaret Latham, Higher Clerical Officer to the Newcastle Haemophilia Centre, for her considerable help in the organisation of this presentation. They would also like to thank the Newcastle Area Health Authority, the Northern Regional Health Authority, and the British Haemophilia Society for their continued help and financial support, both with this presentation and with the day-to-day work of the Centre. Much help has also been given by members of the Department of Haematology, and by the University Department of Photography and Teaching Aids Laboratory.

Rev.
Alan Tanner I would like to express, on the audiences behalf, very warm thanks to Dr Peter Jones and the Newcastle team.

I think numerically the people on the platform now, together with those who took part in the first half of the afternoon, just about makes up a football team! But they obviously have other gifts as well and we are very grateful for them having given us such a splendid session and such a fine start to the Congress today.