SOCIAL AND BEHAVIOURAL PATTERNS IN HAEMOPHILIC CHILDREN AND THEIR FAMILIES AS SEEN AT KENYATTA NATIONAL HOSPITAL, NAIROBI, KENYA

BY

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F E B R U A R Y 1 9 8 5

D E C L A R A T I O N:

I hereby certify that this Dissertation is my own original work and has not been presented for a Degree in any other University.

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Social and behavioural patterns of 38 haemophilic children and their families, attending Haematology Clinic of Kenyatta National Hospital between March and July 1984 are presented.

Haemophilia A accounted for 33 (86.8%), Christmas Disease 4 (10.5%) and Von Willebrand's Disease 1 (2.6%) of all the cases.

17 out of 30 parents interviewed were worried, 9 were afraid while 4 got confused at the first bleeding episode of their child. The feelings of the patients (only those aged 5 years and above) regarding their illness were varied. 25 (78.1%) were unhappy, 6 (18.8%) were afraid of another bleeding episode and 1 (3.1%) was ashamed of his condition. It was noted that as the haemophilic child grew older, he developed satisfactory adaptation towards his illness.

All the mothers interviewed wondered what sins they had committed before or during their marriage to suffer such affliction in the family. All the fathers on the other hand, expressed relief that they were not involved genetically in the transmission of the disease. Only 7 (23.3%) parents thought that their families had been bewitched. 26 (86.7%) of the healthy siblings felt sympathetic for their sick brother, 3 (10.0%) felt indifferent while only 1 (3.3%) felt that the sick brother was a bother.

This study emphasises the need of continuous emotional supportive care from the physicians.

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INTRODUCTION

Haemophilia is a sex-linked recessive bleeding disorder for which there is no cure. However, medical advances resulting from isolation of individual clotting factor concentrates of plasma fractions have made it possible to control excessive bleeding more effectively than in the past (1).

Taylor, Ahluwalia, Morrison, Kaviti and Lucille Cardwell (2) in 1968 noted that very few cases of Haemophilia A were recorded among the Africans in Kenyatta National Hospital (K.N.H.) records. In their study which they carried out in the same year at K.N.H. they, however, saw seventeen cases of Haemophilia A within a period of less than a year. Much earlier in 1966, Forbes, Mackay and Khan (3), in their study at K.N.H. had described the clinical features of five patients with classical Haemophilia A and four with Christmas Disease. These workers reviewed the literature on Haemophilia in Africa as well and noted that although there were a few recorded cases, it was probably commoner in East Africa than was realised. They suggested that adverse environmental factor including ritual circumcision reduced the number of the affected males. Lothe (4) in 1968 described eight Ugandan patients seen between 1959 and 1967.

Kasili and Kariithi (5), in their study of fourty-two patients in 1978 at K.N.H. found that Haemophilia A was the single most common haemorrhagic disorder accounting for 50.2%, Christmas Disease was the runner-up (26.2%), but it was diagnosed more frequently in this series than in the previous series from Kenya and elsewhere (6) but not as high as reflected in Forbes et al (3). Von Willebrand's Disease

which had not been documented in any of the African series reported before, contributed about 18% of their cases. Later, Kitonyi and Kasili (7) carried out a retrospective study of one hundred and five patients with hereditary haemorrhagic disorders seen at K.N.H. between March 1975 and July 1980 and found that Haemophilia A formed 63% of all the cases, Christmas Disease, 22% and Von Willebrand's Disease, 9.5%. In these two studies all ethnic groups were represented in the hereditary haemorrhagic disorders. There were 2 subjects of Asian descent included in Kasili et al. (5) study while three subjects of Asian descent were included in Kitonyi et al (7). The high incidence amongst the Kikuyu noted in both studies probably reflects the fact that most Kikuyus relatively live nearer K.N.H. and have easy accessibility to facilities offered at K.N.H. than most other tribes.

Although tribal distribution has been studied, social and behavioural patterns have not previously been looked at in Kenya.

Despite the ease of diagnosis of Haemophilia, availability of therapeutic procedures, Haemophilia is poorly understood with regard to long term emotional needs imposed by it. This probably accounts for the paucity of literature on social and behavioural patterns of haemophiliacs.

Most patients with Haemophilia show an onset of symptoms in early childhood and are subjected to repeated bleeding episodes, often causing severe pain and requiring immobilization, hospital admissions and various treatment procedures. Thus, a constant threat of fresh bleeding looms over the haemophilic child.

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Intimately related to this is the feeling of guilt which the patient's mother has to bear because she is the carrier of the disease. These maternal guilt feelings may be aggravated by the father's rejection of the child or by the father's lack of cooperation in child-rearing. Worse still is the reluctance of many physicians to undertake the long term continuous care of the young haemophilic patient.

Browne, Malley and Kane (8) in their two-year study (between 1957 and 1959) of twenty-eight haemophilic children and their families at the University of Pittsburgh, Pennsylvania, U.S.A. found that emotional factors contributed to the timing of spontaneous bleeding episodes in Haemophilia. This was confirmed by Mattsson and Gross (9, 10) in their study of thirty-five haemophilic boys and their parents who were seen in psychiatric interviews over a period of 2 years (between 1963 and 1965) at Columbus, Ohio, U.S.A. This study further showed that clinical manifestations of the disease do not always correspond to the expectations based on the level of the clotting factors. Stress in a haemophilic child for example, was noted to affect blood vessels, the amount and frequency of bleeding. An example to this was the spontaneous bleeding encountered in 8 patients, 4 of whom were brothers (Mattsson et al (9)). These patients often began to bleed, without previous trauma, just prior to a highly anticipated event, for example, a weekend camping.

These workers (8, 9, 10) noted that the haemophilic children were usually passive although they would rebel in subtle ways against restrictions placed upon them. It was also noted by these workers that the mothers felt intensely guilty when told that they were carriers of the disease. As a group, the

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mothers were depressed, anxious women who tearfully discussed their haemophilic children. A few saw the haemophilic child as a cross to bear. Majority of the fathers, on the other hand, not withstanding their feelings of apprehension, took a decisive active part in caring for the patients from the time of diagnosis. Family intergration and adaptation to the task of raising a haemophilic child were better in the families where the father had taken active part in caring for their haemophilic children than in those families where the fathers were away or had evinced uncooperative attitudes.

Another interesting observation noted by these workers was the active participation of the older healthy siblings in the care of the bleeder. These older healthy siblings assumed a protective role both at home and away.

In all the studies on Haemophilia and its allied conditions at K.N.H., Kenya and elsewhere in Africa, no attempt has been made to study the social and behavioural patterns of these patients and their families hence the impetus for this study.

OBJECTIVES

The objectives of this study are:-

(1) To determine the socio-economic status of the families of haemophilic children.

(2) To study the behavioural pattern of these patients and their families regarding:-

(i) acceptance of the disease, (ii) adaptation to the disease, (iii) adaptation and acceptance of therapy, (iv) interactions between the patient, the family members and other concerned people.

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With the above objectives the following aims are anticipated to be achieved:-

(1) To provide data as a basis for continuing evaluation of the haemophilic child as he relates to his disease and to his life situation in general.

(2) To provide an insight into the relationship between family attitudes and the patient's clinical course and personality development.

(3) To provide information regarding a rational approach to the management and support of the haemophilic child.

MATERIALS AND METHODS

The Study Period and the Area of Study:-

The study was carried out between March and July 1984 in the Haematology Clinic and Paediatric Wards of Kenyatta National Hospital.

Patients:-

All haemophilic children aged 1 to 18 years attending the Haematology Clinic as well as those admitted directly to the Paediatric Wards were admitted into the study on the basis of their availability at the time the study was being undertaken.

For each patient, the following information was obtained and recorded on the Appendix I.

(a) Particulars of name, age, sex, tribe, race, residence, age (date) at first episode of bleeding and subsequent bleeding episodes. The cause of the bleeding as well as the site of the bleeding.

(b) Physical examination was carried out either at first presentation or on the first contact with the patient.

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The following were noted during the physical examination:-

General condition of the patient, degree of pallor, temperature, evidence of bleeding tendency and whether there was any swelling and or deformity of the joints. Any other abnormal presentation seen was also noted.

Parents and or guardians of these haemophilic children and the patients whose age was 5 years and above were interviewed repeatedly using the questionnaire (Appendix II) and their behaviour observed during their stay in the Paediatric Wards and in the Haematology Clinic.

The social and behavioural background of the patients and their families including family history of bleeders were enquired into using the same questionnaire as illustrated in Appendix II. This questionnaire was designed with the help of all the Social-workers attached to Kenyatta National Hospital and its units, and was pretested both in the Paediatric Wards and in the Haematology Clinic prior to the study.

Classification of the families in the study into different income groups was done with the help of the staff of Central Bureau of Statistics, Ministry of Finance and Planning, Republic of Kenya. (Appendix III).

The socio-economic strata is as shown:-

(1) Lower income group comprises persons with monthly earnings below KSh.699/=.

(2) Middle income group comprises persons with montly earnings between KSh.700/= - KSh.2,499/=.

(3) Upper income group comprises persons with monthly earnings of KSh.2,500/= and above.

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Laboratory Investigations:-

(1) A 2 ml. sample of venous blood was collected into a sequestrene bottle by the author for the following tests:-

(a) Full blood count using the Coulter Counter Model "S" in the Haematology Laboratory.

(b) A peripheral film stained with May-Grunwald Giemsa was routinely reported by competent Laboratory Technicians in the Haematology Laboratory.

These results were used to rule out anaemia and malaria.-

(2) Coagulation Screen tests (Bleeding time, Prothrombin time. thrombin time, kaolin cephalin clotting time as well as factors VIII and IX assays - Appendix IV) were done on all the new patients using Standard Laboratory techniques applied in the Haematology Laboratory, whereas for the old patients the results of Coagulation Screen at first presentation were obtained from the patients' records. The practical technical details followed are as detailed by Dacie and Lewis (11).

Haemophilic patients were graded according to WHO (6) recommendation for grading Factors VIII and IX activity as will be referred to in Table VI, that is,

Less than 1%	=	Severe Haemophilia
1 - 5%	=	Moderately severe Haemophilia
5 -20%	=	Mild Haemophilia

R E S U L T S

The number of patients admitted into the study were 38 representing 34 families. Of these, 36 were males and 2 females.

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One female had Von Willebrand's Disease and the other female, Christmas Disease. Frequency distribution of various haemorrhagic disorders in the study is as shown in Table I. Haemophilia A accounted for 33 (86.8%), Christmas Disease 4 (10.5%) and Von Willebrand's Disease 1 (2.6%) of all the cases.

Table I: Frequency distribution of various haemorrhagic disorders in the study:

Disorder	Number of patients	Percentages
Haemophilia A	33	86.8
Christmas Disease	4	10.5
Von Willebrand's Disease	1	2.6

Referral patients were 30 constituting 78.9% of all the cases. Of these, 25 came from Provincial/District Hospitals and the rest came from City Council Clinics in and around Nairobi. The remaining patients came directly to Kenyatta National Hospital without being referred.

The age distribution of the patients in the study ranged from 14 months to 18 years (Table II). 8 patients were below 5 years of age and the remaining 30 were 5 years or above.

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Table II: Age distribution of the patients in the study:

Age at time of study in years	Number of patients	Percentages
0 - 4	8	21.1
5 - 9	19	50.0
10 - 14	10	26.3
15+	1	2.6
Total	38	100.0

Of the 38 patients, 30 were of school age. However, only 25 were attending school, three among them were in Nursery Schools, 21 in Primary Schools and only one was in a Secondary School. The school attendance in all the cases was irregular.

More than half of 34 families (53.8%) represented in the study fell in the lower income group. The fathers of two other families (5.9%) held professional or executive positions while the remaining 13 families (40.3%) were in the middle income group.

In ten families one or both parents had attained secondary school education level. In other five families none of the parents had any formal education while the remaining nineteen families, one or both parents had attained primary school education level.

Thirty-three families had a Christian background while only one family had a Muslim background.

Tribal distribution of various haemorrhagic disorders is as shown in Table III. The Kikuyu predominated making 57.9% of total number of cases in the study.

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Table III: Tribal distribution of haemorrhagic disorders in the study:

Tribe	Haemophilia A	Christmas disease	Von Willebrand's disease	Nos. of patients	Percentages
Kikuyu	20	1	1	22	57.9
Kamba	3	0	0	3	7.9
Luo	2	0	0	2	5.3
Luhya	1	1	0	2	5.3
Kalenjin	1	1	0	2	5.3
Meru	2	1	0	3	7.9
Embu	2	0	0	2	5.3
Kisii	1	0	0	1	2.6
Somali	1	0	0	1	2.6
Total	33	4	1	38	100.0

Table IV shows the distribution of the patients by district of origin. The parents of one patient included in those of Kiambu District had migrated from Muranga District.

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Table IV: Distribution of the patients in the study by District of origin:

District	Number of patients	Percentages
Elgeyo Marakwet	1	2.6
Embu	2	5.3
Kakamega	2	5.3
Kiambu	10	26.3
Kirinyaga	1	2.6
Kisii	1	2.6
Machakos	3	7.9
Marsabit	1	2.6
Meru	3	7.9
Muranga	10	26.3
Nandi	1	2.6
Nyeri	1	2.6
South Nyanza	1	2.6
Total	38	100.0

Relevant Family History of Bleeders:-

24 out of 34 families in the study had a known family history of bleeders. However, only 16 families volunteered the information that they had more than one bleeder in the family line. In none of these families did the mothers have prior knowledge about the possible mode of inheritance of the disease as young adults and had therefore not informed their husbands about it before their marriages.

Diagnosis was made in 30 out of the 38 patients (78.9%) before the age of 5 years. The remaining 8 (21.1%) patients had diagnosis of haemorrhagic disorder made at 5 years of age or after (Table V).

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Table V: The Ages of the Haemophilic Patients (N = 38) at diagnosis:

Age at diagnosis in years	Number of patients	Percentages
0 - 4	30	78.9
5 - 9	7	18.4
10 - 14	1	2.6
15+	0	0

The age at diagnosis of haemorrhagic disorder was inversely related to the severity of the disorder (Table VI).

Table VI: The Age at diagnosis and Severity of Haemophilia A alone as assessed by Factor VIII activity (N = 33)

Age at diagnosis in years	Factor VIII activity				Nos. of patients	Percentages
	Less than 1%	1-5%	5-20%	Assay not done		
0 - 4	16	5	4	1	26	78.8
5 - 9	4	2	0	0	6	18.2
10 - 14	0	1	0	0	1	3.0
15+	0	0	0	0	0	0.0
Total	20	8	4	1	33	100.0

The number of bleeding episodes was greater in the younger patients with a trend towards fewer bleeding episodes as the patient grew older (Table VII).

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Table VII: Bleeding episodes in relation to age of the patient:

Age at time of study in years	Number of patients	Average bleeding episodes per patient per year
0 - 4	8	10 or more
5 - 9	19	5 to 10
10 - 14	10	2 to 4
15+	1	0 to 1

Clinical Presentation:

The clinical presentation in majority of the patients is indicated in Table VIII. Haemarthrosis was the single most frequent clinical presentation as it accounted for about 93% of all the patients. The runner-up was bleeding due to a haemostatic challenge. Rare presentation included persistent generalised lymphadenopathy and neurological symptoms probably due to intracranial haemorrhage.

Some of these presenting features occurred in combination in the same patient.

It is worth noting that these clinical presentations occurred mainly as spontaneous bleeding episodes except for the bleeding from cut-wounds, tooth extraction and or following circumcision.

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Table VIII: Clinical Presentation in majority of the Haemophilic Patients:

Presentation	Number of patients	Percentages
Haemarthrosis	35	92.6
Excessive bleeding from cut-wounds/ Tooth Extraction/ Circumcision	17	44.7
Haematoma/Ecchymosis	12	31.6
Epistaxis	6	15.6
Bleeding from gums	4	10.5
Bloody diarrhoea	2	5.3
Hemiplegia	2	5.3
Haemetemesis	1	2.6

Reaction of the Parents to the first bleeding episode:

Majority of the parents (56.7%) were worried, 9 (30%) were afraid and 4 (13.3%) became confused at the first bleeding episode in their child (Table IX).

Immediate medical attention was sought by the parents once they realised that their child had a bleeding problem.- However, according to the volunteered information, none of the parents conceded to having taken their child to a traditional doctor (healer).

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Table IX: Emotional reaction to the first bleeding episode by the accompanying parents:

Reactions	No. of mothers	No. of fathers	Total Nos. of parents	Percentages
Being afraid	6	3	9	30.0
Being worried	12	5	17	56.7
Being confused	2	2	4	13.3
Total	20	10	30	100.0

Accompanying Persons to the Clinic/Wards:

Twenty (52.6%) patients were accompanied by their mothers, ten (26.3%) by their fathers and five had guardians accompanying them while three (7.9%) were unaccompanied.

The mother was the most involved in taking the patient (her son) to the hospital during subsequent bleeding episodes. The father, however, was involved in restricting the movements or play and other activities of the child (his son).

The feelings elicited in the parents regarding the chronic nature of the illness were varied as shown in Table X. Only 7 out of 30 parents felt that their family had been bewitched.

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Table X: Psychological feelings, thoughts and beliefs about the problem as a chronic recurrent illness

Feelings	No. of mothers	No. of fathers	Total Nos. of parents	Percentages
The family has been bewitched	5	2	7	23.3
The family has been cursed	1	0	1	3.3
These are temptations (trials)	10	3	13	43.3
Never thought of anything	5	5	10	33.3

All parents with only one exception did not blame the recurrence of the bleeding on inadequate medical treatment. Twenty-five (83.3%) of the accompanying parents conceded to having no explanation concerning their ill-children having been given by a doctor. The remaining five (16.7%) agreed that they had had some explanation although this was unsatisfactory as far as they were concerned and requested for more explanation.

Most of the healthy siblings (86.7%) particularly those who could understand the problem, felt sympathetic for their sick brother (Table XI) and the older healthy siblings took active part in caring for their sick brother.

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Table XI: Feelings of other children (siblings) towards the child with the bleeding disorder (N=30)

Feelings	No. of children (Siblings)	Percentages
Felt sympathetic	26	86.7
Felt that he is a bother	1	3.3
Felt indifferent	3	10.0

The feelings of the patients (aged 5 years and above) regarding their illness is as illustrated in Table XII. One of the patients was both ashamed and unhappy about his illness while two patients felt both unhappy and were afraid of having another bleeding episode. Six (18.8%) patients had a constant threat of another bleeding episode looming over them. 25 (78.1%) were totally unhappy about their illness.

Table XII: Emotional Feelings expressed by the Haemophilic Patients (N = 32)

Feelings	Number of patients	Percentages
Feeling ashamed	1	3.1
Feeling afraid of another bleeding episode	6	18.8
Feeling unhappy	25	78.1

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D I S C U S S I O N

All ethnic groups are not represented in this study. This may be because the period of study was short and hence not all the 172 documented haemophilic patients could avail themselves for the study. Again other constraints like scarcity of money to pay for the bus-fare to Nairobi may have contributed to this just as the results show that majority of the patients came from families of low income group.

The large number of Kikuyus in the study probably reflects the fact that most Kikuyus live in areas close to K.N.H. as compared with other tribes. This compares very well with the findings in Kasili et al. and Kitonyi et al. studies (5, 7).

In this study it was found that all the mothers who were born in families with clear family history of bleeders had no knowledge at all about the mode of inheritance of the disease as compared to those mothers in U.S.A. (8, 9, 10). The reason for this difference is lack of exposure to medical knowledge in our environment.

It is worth noting that the age of diagnosis of haemophilia was inversely related to the severity of the disorder. This is in keeping with what is expected of all inherited blood clotting disorders (6).

Mattsson et al. (9) found that there was a diminution in bleeding episodes as the haemophilic child grows older. This compares well with the finding in this study. This clinical improvement of most older haemophilic patients has been attributed to their greater cautiousness, the protection of a well-developed musculature and other, as yet ill-defined factors related to the complex biological changes of Adolescence.

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Haemarthrosis was the single most clinical presentation seen in this study. This again compares well with the findings in Kasili et al. and Kitonyi et al. studies (5, 7).

Most presenting clinical features seen in the patients in this study occurred mainly as spontaneous bleeding episodes without a haemostatic challenge. This is in keeping with the findings in Browne et al. study (8). It is thought that the timing of spontaneous bleeding episodes in haemophilia may be related to emotional factors. The specific emotional conflict which was noted in the patients in this study and which compares well with other studies (8, 9, 10) involved the desire to be active and aggressive like any other normal child and the inhibition of this wish. The inhibition in the patient is a reflection of father's restriction and the mother's anxiety about the patient's activity and the harm to which she feels it will lead. Being passive becomes equivalent of being good and pleasing the mother. Bleeding also brings passivity. Passivity is further encouraged in the child by mother's overprotective attitude towards him coupled with rigid precautions or restrictions carried out by the father.

In the study it was noted that the mother was the most involved in taking the patient to the hospital during subsequent bleeding episodes. This was in agreement with an observation made by other workers (8, 9, 10). Perhaps out of guilt as the carrier of the disease, the mother takes full responsibility for the care and management of the child. In contrast, it is known for example that in families with a leukaemic child or a child with other disease conditions, the fathers are quite active in seeking help for the child.

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The study shows that most of the siblings (86.7%), particularly those who could understand the problem, felt sympathetic for their sick brother and the older healthy siblings took active part in caring for their sick brother. This is in keeping with the findings of Mattsson et al. study (9).

As noted by Browne et al. (8), haemophilic children and their parents are faced with many similar problems peculiar to haemophilia and they often show a rather characteristic sequestial pattern in coping with these problems. The behaviour of both the young bleeder and his parents is therefore, generally predictable. The observations in the present study bear out the importance of these factors.

Observations on the Patients:-

Most of the children (65.8%) were outwardly passive, however, they would rebel in subtle ways against the restrictions placed upon them. They would use their vulnerability to manipulate situations. Common threats were: "If you beat me, I'll bleed", "If you punish me, I'll tell the Doctor".

It was generally observed that as the patients (bleeders) grew older they showed a satisfactory to optimum adaptation to their illness. They thus function effectively at home, in school, at work with their peers with a few or no limitations other than those realistically imposed by the disease and its complications e.g. hemiplegia.

The patients below 5 years had past bleeding episodes which were associated with "bumps" or "falls". Those above 5 years had increasing comprehension of relationship between physical trauma, bleeding and the common treatment procedures. Their better understanding of the necessary physical restrictions

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together with a widening acceptance of compensatory activities such as reading, playing with less dangerous objects helped them cope with repeated haemorrhagic episodes.

In the younger children, anxiety caused by pain, immobilizations and the separation from the parents at the time of admission was apparent. The older children in addition displayed anger and impatience when admitted mixed with feelings of sadness and hopelessness about the chronicity of their disease.

Reactions of the Parents to the diagnosis:-

In general, the mothers interviewed felt intensely guilty when the diagnosis was explained to them and were told that they were the carriers of the disease. This initial reaction, however, was not affected by variables such as age of the child at the time of diagnosis, severity of the disease, presence of other bleeders in the family and the education background of the mother. Most of them wondered what they had done before or during their marriage to bring this affliction upon themselves. None of them, however, did have prior knowledge of the possible mode of inheritance of the disease as young adults before their marriage. In contrast to the guilt feelings of the mothers, the reaction of the fathers to the diagnosis was varied. Most of them were anxious and many expressed relief that they were not involved genetically. However, two fathers remained aloof and remote and did not want to be associated with the sick children.

Adaptive behaviour to cope with the situation by the parents:-

Almost all the mothers (95.0%) had learnt to cope with the initial reaction of anxiety,sadness and guilt, following the explanation of the diagnosis of the bleeding disorder with

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only one exception. This particular mother showed a strong reaction of anger and guilt from the time she learnt of the diagnosis and refused further interview during subsequent visits to the Haematology Clinic.

The successful adaptation to the task of raising a bleeder seen in these mothers did not seem to be influenced by such variables as the child's age at diagnosis, the clinical severity, the presence of another bleeder in the family or the education background of the mother.

Fathers cooperated in taking care of their ill-sons, but employed rigid precautions in bringing them up. They tried to relieve the guilt of their wives by stressing their mutual responsibility for having borne a bleeder and for raising him. Only two fathers, mentioned earlier, had evinced uncooperative attitudes following the diagnosis. One of them showed obvious hostility to his wife and was planning to marry another wife.

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C O N C L U S I O N

It was found that the majority of the patients in this study came from the low income bracket. This is in keeping with the fact that K.N.H. serves mainly low income patients.

The study shows that majority of the haemophilic children would rebel in subtle way against the restrictions placed upon them. They would use their vulnerability to manipulate situations. This is the experience of other workers too (8, 9, 10). Continuous counselling and support by the physician is therefore essential for emotional growth of the young haemophilic patient as it is for his state of physical well-being.

It is also evident from the study that the majority of the mothers interviewed could cope up with the initial reaction of anxiety, sadness and guilt, following the explanation of the diagnosis of the bleeding disorder. It is, therefore, important that once the diagnosis is made, the physician must be capable of supplying the parents with comprehensive and repeated information about the illness, its mode of inheritance and therapeutic concepts. On the other hand, he must exercise restraint regarding counselling about pregnancies.

The study also showed that by the age of 5 or 6 years, the haemophilic child begins to comprehend the nature of his illness and at this time he should be given some responsibility for cooperation in his care and protection.

Most of the siblings particularly those who could understand the problem were shown to be sympathetic and the older ones took active part in caring for their haemophilic brother. Hence the older sibling should be actively involved in the care of the haemophilic brother.

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Finally, the parents should, from the very beginning be supported by the physician in raising their haemophilic children. This may be achieved by promoting reasonable play activities with other children and by discouraging undue restriction and protection.

R E C O M M E N D A T I O N S

(1) Health education of the parents as regards the nature, aetiology, management, disease course, prognosis, and new medical concepts of the disease should be intensified.

(2) The need of continuous emotional supportive care from the social workers, nurses, and the physicians should be emphasised. Questions raised by the parents, the haemophilic child and his siblings about the hereditary nature of the illness must be answered in an honest and realistic manner.

(3) The ongoing emotional support to the parents should also stress the need for their active participation in the care of their haemophilic offspring.

(4) The haemophilic child who is able to comprehend the nature of his illness should be given some responsibility for cooperation in his care and protection.

(5) There should be a complete program of necessary immunizations and proper dental hygiene for the haemophilic child.

(6) Orthopaedic Surgeon, the attending physician (family physician) deserve and often require counselling by the Paediatrician, or Physician experienced in the treatment of bleeding disorders.

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APPENDIX I

Name ..... Age ..... Sex .....
Tribe ..... Residence .....
Race ..... Referred .....
Date of admissions: 1st .....
2nd .....
3rd .....
etc .....
Presenting complaints (specify) Duration .....
.....
Related/Unrelated .....
Past Medical history (specify) .....
.....
Cause of previous admissions .....
.....
Previous treatment (+ duration) .....
.....
Transfusions (number) .....
Transfused with what .....
Operations if any .....
Special/Family history (relevant) .....
.....
Siblings' health .....

CLINICAL ASSESSMENT

General condition (specify) .....
Pallor .....
Temperature ..... Oedema .....
Nails ..... Tongue .....
Conjunctivae .....
Any evidence of bleeding .....
Site of bleeding .....
Any evidence of joint deformity/swelling .....
.....

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APPENDIX II

PART I

Code ..... Sex ..... Age .....

Tribe ..... Race .....

Religion: Christian ..... Muslim .....

Others (specify): Level of education .....

School attendance: Regular/Irregular .....

PARENTS:

Marital status: Single ..... Married.....

Separated/divorced .....

Widow/Widower .....

Occupation: Employed ..... Self-employed .....

..... Unemployed .....

Others (specify) .....

Monthly earnings: Father ..... Mother .....

Level of education: Std. 1 - III) Primary
Std. IV - VII)
Form I - IV
Form V - VI
University and above
Not at all.

PART II

(To be answered by the patient)

1) Do you have any illness which makes you come to hospital?
   (a) Yes (b) No
2) If the answer to the question is "a" do you bleed excessively when you cut or injure yourself? (a) Yes (b) No
3) If the answer to question 2 is "a" when did you start bleeding excessively for the first time?
   And what happened?
   (a) Taken to hospital at once
   (b) Contacted a local medicine-man
   (c) Nothing was done until some hours passed
   (d) Not taken to hospital.
4) Who took you to hospital or to medicine-man (traditional doctor) when you first bled? (a) Mother (b) Father (c) Brother (d) Sister (e) Others (specify)
5) Who takes you to hospital whenever you are hurt or bleed? (a) Mother (b) Father (c) Brother (d) Sister (e) Others (specify)

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6) Have you ever been admitted to the hospital as a result of bleeding? (a) Yes/No (b) How many times?

7) Does your illness affect (limit) your relations to your parents' siblings, opposite sex and peer groups? (a) Yes/No (b) Specify the most affected.

8) Are you restricted on your movements/play as a result of your illness? (a) Yes/No (b) Who restricts you most? (i) Father (ii) Mother (iii) Others (specify)

9) What are your feelings about your illness? (a) Ashamed (b) Afraid (c) Unhappy (d) Others (specify)

10) Does your illness affect your (a) Work (b) School attendance (c) Social interactions (d) Hobby/play (e) Not at all

11) If the answer to question 10 is either "a", "b", "c" "d" or "e" give brief explanation.

PART III

(To be answered by the Parents/Guardians)

1) Who are you to the sick? (a) Parent (b) Guardian

2) How many children do you have? .....

3) Which number in the line of the siblings does the sick child fall? .....

4) Do you have any other child/children (a) your family (b) your brother's family (c) your wife's/husband's (d) your wife's family/husband's family with a similar illness?

5) What was your reaction (feelings) when this child got affected by this illness? (a) Afraid (b) Worried (c) Confused (d) None of the above.

6) What steps did you take? (a) took the child to hospital (b) took the child to a traditional doctor (c) gave a local first aid (d) left him unattended.

7) Who has been taking the child to the hospital whenever this has recurred (happened)? (a) Mother (b) Father (c) Brother (d) Sister (e) Neighbours.

8) If this illness has been recurring what has been your feeling about it? (a) Your family has been bewitched (b) Your family has been cursed (c) These are temptations (d) Never thought of anything.

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9) If the answer to question 8 is neither 'a' nor 'b' nor 'c' then do you think the recurrence has been caused by inadequate medical care? (a) Yes (b) No

10) Have you ever been explained by the doctor what your child is suffering from? (a) Yes (b) No
And have you been aware of such problem occurring in your family long before you got married (applies to the mother only)

11) If the answer to question 10 is Yes what are your feelings about it.
(a) Too bad for the child
(b) Divorce or separate with my wife/husband
(c) Re-marry someone else/allow my husband to marry someone else.
(c) Neglect the child
(e) Leave the responsibility of taking care of the child to the mother
(f) Accept the situation as it is.

12) Give a brief explanation to your answer of choice in question 11 .....
.....

13) As the mother of the sick child, how do you feel after you have known the cause of the illness of your child?
(a) Not worthy living together
(b) Feel unworthy
(c) Not to bear any more child
(d) Not affected because I am not the only one.

14) As a married person, what is your partner's feelings towards this child's illness?
(a) Not worthy living together
(b) Child to be fully hospitalized
(c) Wife to be treated in order to stay together
(d) Not to have any other child in future.

15) As a parent/guardian, what do you think about the future of this child? (a) Care for the child
(b) Government should come in and help
(c) Will abandon him in hospital.

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16) Has your child's illness affected the economic status of your family? Yes/No. If yes, how.

17) What is the reaction of other children towards this child's illness?
(a) Feel sympathetic
(b) Feel that he is a bother
(c) Feel indifferent

18) Has this child's illness compromized or affected your love/your husband's love towards other children? Yes/No.

19) Give any of your comments about the behaviour and management of this type of children (haemophilic) while at home .....
.....
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Thank you.

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APPENDIX III

NAIROBI CONSUMER PRICE LIST - CENTRAL BUREAU OF STATISTICS, MINISTRY OF FINANCE AND PLANNING, REPUBLIC OF KENYA.

Base: January - June 1975 = 100

JUNE 1984

COMMODITY GROUP	INCOME GROUP		
	LOWER	MIDDLE	UPPER
Food	279.1	266.9	282.4
Drink and Tobacco	300.0	272.8	314.7
Clothing and footwear	384.2	254.2	268.7
Rent	364.6	331.6	300.5
Fuel and power	406.5	378.0	343.2
Furniture, furnishing, household equipment and household operation	340.8	328.5	298.4
Health and personal care	236.3	314.8	417.0
Transport and communications	289.7	399.5	323.8
Recreation, entertainment and education	159.6	208.9	226.4
Miscellaneous goods and services	315.9	255.7	317.1
Average weighted index for all groups	314.1	294.4	300.1
% change from June 1983 to date	10.4	8.9	7.4
Average weighted index for all groups excluding rent	299.1	287.3	300.0

FOR THE PURPOSE OF THE INDICES

1. Lower Income Group comprises persons with monthly earnings below KSh.699/=
2. Middle Income Group comprises persons with monthly earnings between KSh.700/= - KSh.2,499/=
3. Upper Income Group comprises persons with monthly earnings of KSh.2,500/= and above.

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APPENDIX IV

ROUTINE COAGULATION SCREEN DONE AT KENYATTA NATIONAL HOSPITAL

	TEST	METHOD
1)	Bleeding time	Ivy's method (Duke's method in infants under 1 year).
2)	Prothrombin time	Quick' One stage (Local Brain thromboplastin standardised with British Comparative Thromboplastin)
3)	Thrombin time	With 6 units/ml (Parkes Davis Thrombin)
4)	Kaolin Cephalin Clotting time (KCCT)	10 minutes incubation done with Diagen or platelin platelet substitute.
5)	Platelet Count	Coulter thrombo counter model "C". Visual estimation always done as well.
6)	Routine haematological determination	Coulter Counter Model "S". Peripheral blood film report.

ADDITIONAL TESTS DONE FOLLOWING COAGULATION SCREENING

	TEST	METHOD
1)	Substitution (correction)	With absorbed plasma, normal plasma and normal serum.
2)	Stypven time	Russel Viper Venom (Orthodiagnostics)
3)	Hess test	As in Dacie (II)
4)	Factor VIII and IX assay	One stage method based on KCCT (DADE substrate plasma).
5)	Factor X assay	Using Russel Viper Venom (Orthodiagnostics).
6)	Factor VII assay	Based on the prothrombin time (Substrate plasma from DADE).

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