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Haemophilia in Zambian Children

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Haemophilia is not as uncommon in Black people as was previously thought (Bullock and Johnson 1957, Gelfand 1945). Several cases of haemophilia have been reported in Africa (Forbes et al 1966, Comperts et al 1969, Robin 1964, Essien and Adeloye 1972). In Zambia, Traug (1970) reported a single case of classical haemophilia in the first issue of the Zambia Paediatric Journal. Twelve cases of classical haemophilia seen during a two-year period (1973-1975) are reported here for the purpose of documentation and to bring to the attention of Medical Practitioners that this disease does occur in the country.

CASE HISTORIES

CASE 1 (S.Z.) This boy was apparently treated at the UTH in 1972 but the records were not available. He was seen for the second time in October 1973 with a swollen tender left thigh and knee associated with pallor but without jaundice. He had hepatosplenomegaly. A provisional diagnosis of Sickle Cell Anaemia was made by the Casualty Officer. 5 days later the knee was more swollen and aspiration of the knee yielded blood. A presumptive diagnosis of traumatic haemarthrosis was made. This was later revised when the child continued to bleed from the puncture site. A diagnosis of Haemophilia was entertained after several blood transfusions had been given. Haematological consultation was requested and coagulation studies were done (see table 1). These were consistent with factor 8 deficiency – classical haemophilia. On this information several maternal male cousins and maternal uncles were screened.

CASE 2 (R.S.) aged 9 months is a cousin of Case 1 and was seen on family screening. The parents gave a history of easy bruisability in the child which had not been taken seriously.

The KPTT performed on Aluminium Hydroxide absorbed plasma with the patient's plasma was 45 seconds. The rest of the studies are shown in the table. These results again were consistent with factor 8 deficiency. The patient's plasma factor 8 concentration was not determined.

The child has since started walking and has had one episode of haemarthrosis successfully treated with cryoprecipitate.

CASE 3 (S.M.) 2 years old, and a cousin of Case 1. The mother gave a history of prolonged bleeding from minor cuts. He was picked up on family screening.

This patient has had two episodes of prolonged bleeding one from a tooth socket, and the second extensive bleeding into the thigh. Both episodes were treated with cryoprecipitate. The KPTT was not markedly prolonged.

CASE 4 (L.M.) was one year when he was seen on family screening. He is also a cousin of Case 1. He had history of easy bruisability. His coagulation tests were compatible with factor 8 deficiency. Quantitative estimation of factor 8 was not done.

This patient was lost to follow up so that his clinical course is now known.

CASE 5 (J.S.) 4½ year old boy, a maternal cousin of Case 1, who had a history of prolonged bleeding and limited extension of the left elbow from previous haemarthrosis in the joint. Coagulation tests proved that he was a haemophiliac.

This boy has had haemarthrosis of the right knee as well as excessive bleeding from a cut lip. Both these episodes were treated with cryoprecipitate and commercial Anti Haemophiliac globulin.

CASE 6 (S.M.) aged 2 years when he was first seen in family screening. He is a cousin of Case 1. He had at that time a history of prolonged bleeding from minor cuts as well as recurrent haematomata from minor trauma to the forehead. His coagulation studies proved that he was a case of factor 8 deficiency.

This boy has subsequently suffered from measles and a haemarthrosis of the right knee. He received cryoprecipitate and commercial Anti-haemophiliac gamma globulin.

The six cases described above are all Tongas from the Pemba area of Zambia.

CASE 7 (K.M.) a 15 year old boy was referred to UTH from Lubwe Mission Hospital, Samfya, with history of prolonged bleeding from minor cuts. He also had bleeding into the joints with limited extension of the right elbow. During a school riot he sustained a cut on his head from a small stone thrown at him. He bled profusely from this and required several units of

TABLE I

	PT	KPTT	KPTT HP + PP (SECS)	KPTT OF NP + HP (SECS)	KPTT NP + PP (SECS)	FACTOR VIII	BLEEDING TIME (MINS) (Normal 1-7)
1	13	61		47	51		3.5
2	13	59					4
3	13	40	43	33	35		N
4	N	59	35	63	49		N
5	N	105	108		70		N
6	N	88	90		66		N
7	N	70				2%	N
8	N	65				2%	N
9	N	70				2%	N
10	N	75		41			N
11	13	61				2%	N
12	14	55					

KPTT - Kaolin partial Thromboplastin Normal = 30-40

PT - Prothrombin Time Normal = 12-15

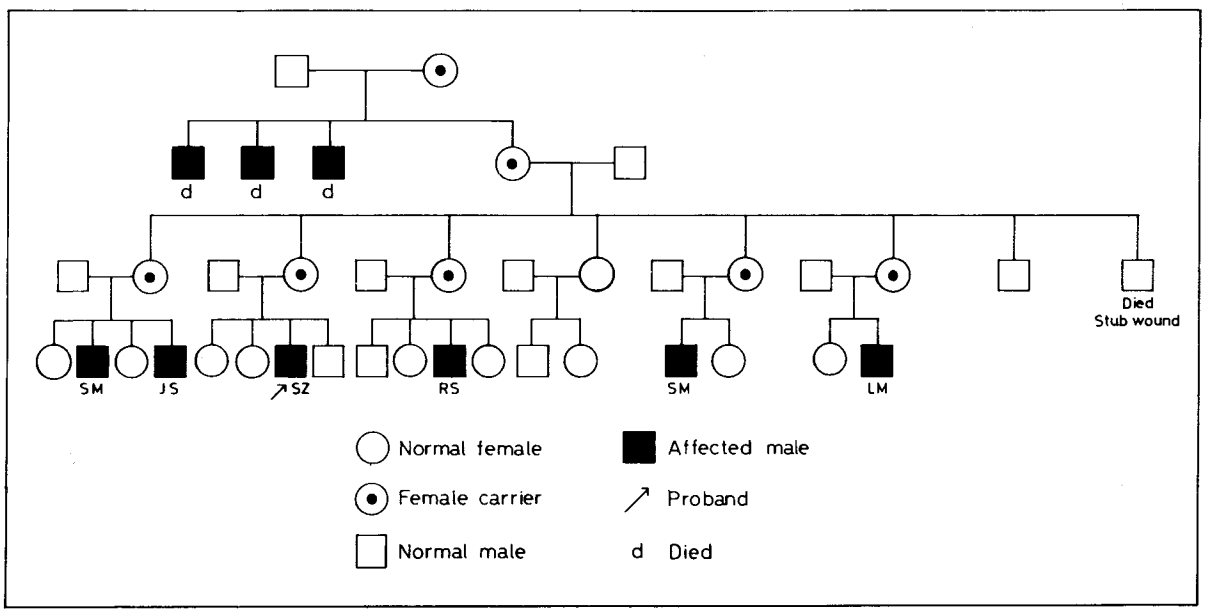
HP - Haemophiliac Plasma

NP - Normal Plasma

PP - Patient Plasma

FIG. I

FAMILY PEDIGREE OF CASES 1-6 HAEMOPHILIACS



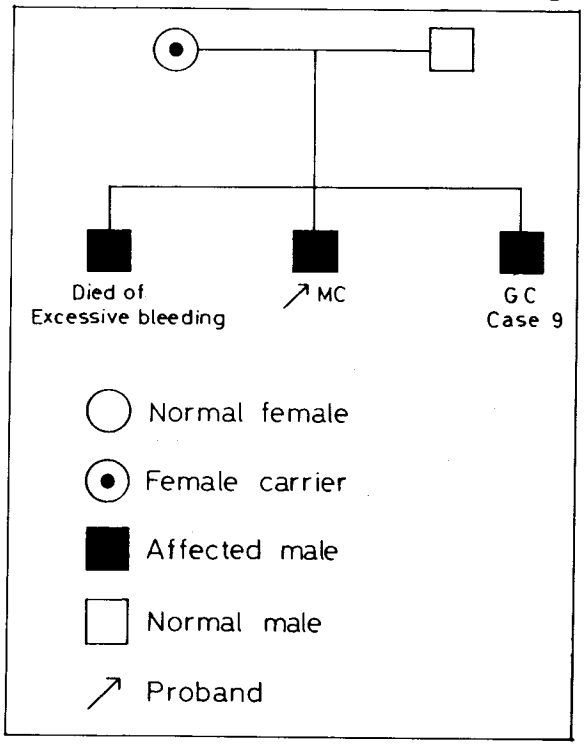
whole blood to arrest the bleeding. He was sent to UTH for investigation. His KPTT was 70 secs as compared to a control of 30 secs. The plasma factor 8 was less than 2%. On further questioning he did admit that 3 of his nephews were bleeders.

The patient has not been followed up in Paediatric Haematology Clinic because of transportation difficulties.

CASE 8 (M.C.) is a 5 year old boy who comes from Pemba (unrelated to the first 6 cases). He was first seen in casualty with a small laceration on a swollen foreskin. A diagnosis of phimosis was made and a circumcision was performed. The child bled profusely from the circumcised area and developed shock. A femoral cut down was done as all the other

FIG. II

PEDIGREE SHOWING HAEMOPHILIAC SIBS OF CASE 8



peripheral sites had been damaged when he required a blood transfusion in another hospital, one or two years ago. Three days after the institution of rescusitative measures, which included an infusion of porcine factor 8 concentrate, coagulation studies were done which showed that he was a haemophiliac, with less than 2% plasma factor 8.

The mother was questioned regarding a bleeding diathesis in other members of the family. One male child died from excessive bleeding and she admitted to easy bruisability in her male infant (Case 9).

CASE 9 (G.C.) 10 months old child brother of case 8 was seen as part of family screening. He had bruises on his forehead and legs. His KPTT was prolonged to 70 secs. The plasma factor 8 was less than 2%. The child developed a haematoma over the venipuncture site. The father refused treatment of the child on the grounds that he believed the child was bewitched during the first few days of his life. This boy and his brother have not been seen for follow-up.

CASE 10 (M.K.) this is a 7 year old Bemba boy who was first seen because of a haemarthrosis of his knee. He has had a history of prolonged KPTT, 75 secs with plasma factor 8 of less than 2%. He was treated with porcine factor 8 as well as cryoprecipitate with good results. Since this initial hospitalization he had two further admissions, one for a fractured right femur and the second for excessive bleeding from a tooth socket. The tooth was extracted without the dentist knowing that he was a haemophiliac. On both occasions he was treated successfully with commercial preparation of human factor 8, cryoprecipitate as well as Episolon Amino Caproic Acid.

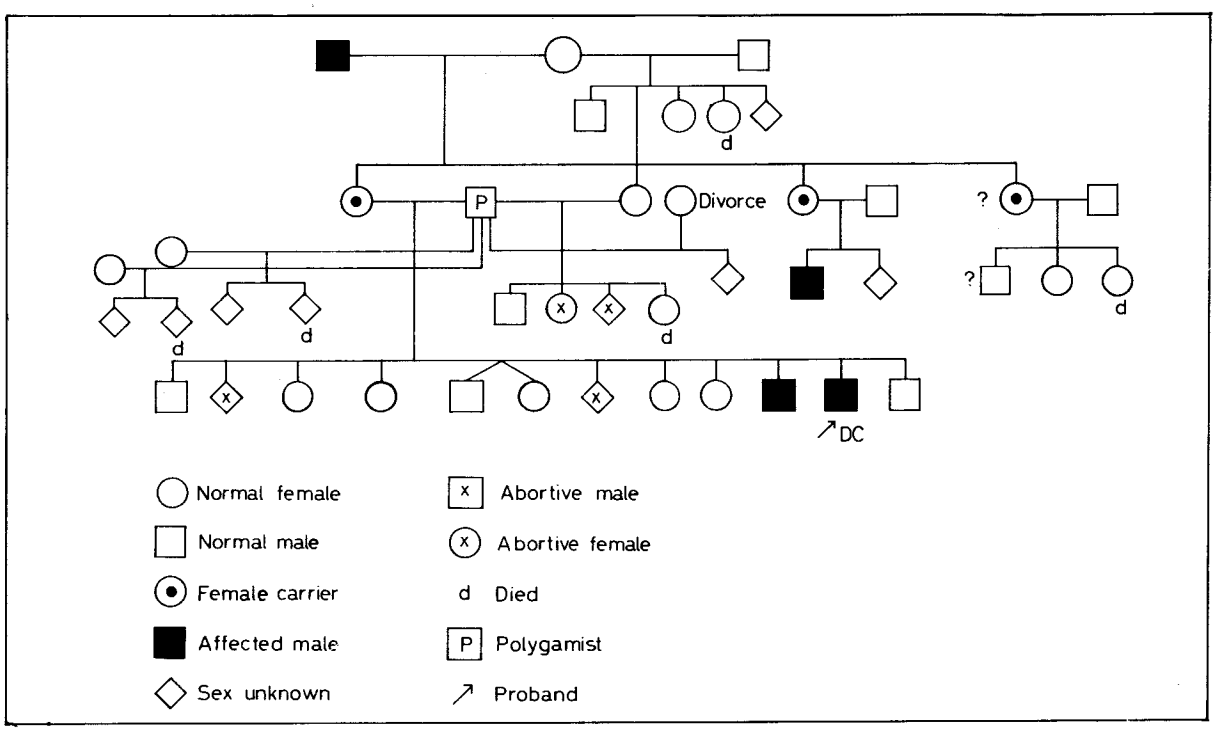
CASE 11 (C.M.) this is a 7 year old Lozi boy who gave a history of excessive bleeding from minor cuts which required several blood transfusions. He was seen in this Hospital in late 1973 because of haemarthrosis. At that time the diagnosis of Haemophilia was made on the basis of an abnormal coagulation test. Since this episode he has had bleeding from carious teeth, haemarthrosis and excessive bleeding from a lacerated lip. He has been treated with porcine factor 8, Episolon Amino Caproic Acid, cryoprecipitate and blood transfusions.

CASE 12 (D.C.) is a 4 year old boy whose Tonga mother is married to a Zimbabwean. He was seen at UTH because of a swollen left knee contracture and wasting of the left thigh muscles. An initial diagnosis of tuberculous arthritis was made on the basis of grade 1 positive Heaf (Heaf grading 0-4) with normal chest X-Ray. A synovial biopsy was made and this was negative for tuberculosis. The patient however bled profusely from the biopsy site and required a blood transfusion. Haemophilia was diagnosed on the basis of a prolonged KPTT and plasma factor 8 of 2%. On further questioning the mother did admit to the child having a bleeding problem since birth and further added that there are several members who bleed easily (see family tree Fig. iii).

DISCUSSION: These cases illustrate the importance of a well taken history which should include past illnesses, familial and hereditary disorders. Unnecessary surgery in a number of these patients could have been avoided and if surgery becomes absolutely necessary the patient should be protected from excessive bleeding by administering factor 8 concentrate prophylactically. In small hospital centres where there are no facilities for doing coagulation studies the history becomes even more important for it will point

FIG. III

PEDIGREE OF HAEMOPHILIACS OF CASE 12 (C.D.)



to a bleeding diathesis and the need to treat any bleeding with fresh whole blood and then to refer the patient to a centre with the essential laboratory facilities for definitive diagnosis.

The importance of family studies cannot be over emphasized. Half of the cases described were diagnosed on family screening. On confirming the diagnosis the parents were given letters which indicated that the children were haemophiliacs and that they should receive fresh whole blood if a transfusion for bleeding became necessary. Advice against intramuscular injections and asprin administration was also given. Because of these letters we have been able to follow up 8 children who were referred back to us when the attending doctors in peripheral hospitals thought they could not control bleeding by simple measures.

The other aspects of family studies are of course genetic counselling and documentation of the number of haemophiliacs in the country. With larger numbers the need for improving our diagnostic and therapeutic facilities will naturally arise.

I have not as yet come across a case of Christmas disease (factor 9 deficiency). This is not as common as factor 8 deficiency but it is my belief that with more accurate surveillance of bleeding disorders it is only a matter of time before it is diagnosed locally.

Christmas disease has already been reported in Kenya (Forbes et al 1966) and in South Africa (Comperts et al 1966).

CONCLUSION

The 12 cases of Haemophilia reported here will, I hope, serve to remind us all to include this disease in our differential diagnosis of bleeding diathesis in Zambian patients.

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