HAEMOPHILIA IN THE BANTU: REPORT OF 67 CASES*

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The incidence and mode of presentation of haemophilia in White races have been well documented.[1-4] The paucity of reports of haemophilia in other ethnic groups has led to the assumption that the disease is not common in Negroes and other non-White races.[5-8] This apparent low incidence may, however, be due to failure to report cases or to lack of statistical data on the incidence of the disease. For this reason it was felt worth while to record the features of 67 Bantu patients with haemophilia diagnosed over a 14-year period.

MATERIAL AND METHODS

The material comprises Bantu patients, or blood samples drawn from Bantu patients, referred to the coagulation department of the South African Institute for Medical Research, Johannesburg, during the period 1954 - 1968. The material was drawn mainly from the Transvaal, but included some cases from the Orange Free State and the Eastern Cape Province. The prothrombin index and thromboplastin generation test were carried out according to standard procedures[9] and the kaolin partial thromboplastin time by the method of Langdell *et al.*[9] The assays for factors VIII and IX were based on the methods of Pitney,[10] and Matchett and Ingram,[11] respectively. During the period 1954 - 1966, the diagnosis of haemophilia was made on the basis of the thromboplastin generation test with correction studies, using plasma from known haemophiliacs and Christmas disease cases. From 1966 onwards assays for factors VIII and IX were carried out on all patients with a prolonged partial thromboplastin time.

RESULTS

During the 14-year study period, 67 cases were diagnosed as classical haemophilia (haemophilia A). Of these, 60 were from the Transvaal. Blood samples submitted from remoter areas were not always accompanied by adequate clinical data, and the results presented are on the data available. All 67 cases were males.

Age Distribution

The youngest patient was an infant of 11 months, and the oldest 55 years. The majority of the cases were diagnosed in childhood, but it is worth noting that no less than 12 cases (18%) were adults (Fig. 1).

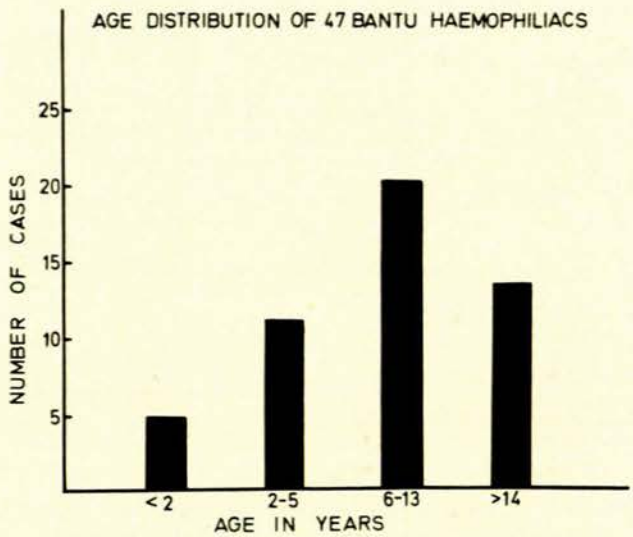

Fig. 1. Age distribution.

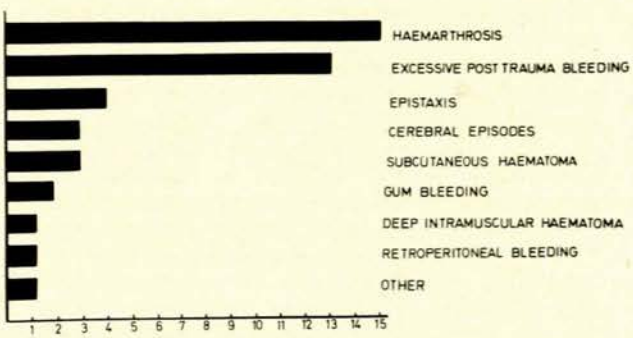

Fig. 2. Clinical features.

Clinical Features

The clinical presentation in 43 cases is summarized in Fig. 2. The most common presenting feature was either haemarthrosis or bleeding after trauma. In 30 of 47 cases with adequate history, there was a previous episode of pathological haemorrhage, and in 10 this necessitated hospital treatment.

Family Studies

These were not carried out in any detail. Two sets of brother-pairs are included in the study, while a history of excessive bleeding in another member of the family was obtained in 17 out of the 47 cases with adequate history.

Factor VIII Levels

Factor VIII was assayed in 21 cases. In 17 patients the level was less than 1% (severe haemophilia) and in the other 4 it was 1 - 5% (moderately severe haemophilia). The severity of the disease was compared with the age at which the patient presented for diagnosis (Table I). No less than 5 out of 17 patients with severe haemophilia presented after the age of 14 years.

During the period when the 67 cases of haemophilia were diagnosed, 8 cases of Christmas disease were seen, i.e. a ratio of 8·4 : 1.

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TABLE I. FACTOR VIII LEVELS IN 21 BANTU HAEMOPHILIACS CORRELATED WITH AGE AT WHICH THE DISEASE WAS FIRST DIAGNOSED

	Number of patients		
Age-group (years)	*Severe haemophilia (factor VIII 1%)*	*Moderate haemophilia (factor VIII 1 - 5%)*	*Mild haemophilia (factor VIII 5 - 25%)*
< 2	1	2	—
2 - 5	4	—	—
6 - 13	5	1	—
> 14	5	1	—
Unknown	2	—	—
Total	17	4	0

DISCUSSION

In 1948 Gelfand[12] noted that haemophilia had never been reported in the Bantu. Shortly after that single case, reports in a Basuto[13] and a Congolese[14] were published. Feldman and Lewis[15] reported on 2 cases in 1952, while Cassel and Metz[16] subsequently documented 8 cases of haemophilia and 1 of Christmas disease seen in Bantu over a 2-year period at Baragwanath Hospital, Johannesburg. Forbes *et al.*[17] reported 5 cases of haemophilia and 5 cases of Christmas disease from Kenya in Kikuyu, Masai and Mchagga ethnic groups, while 2 cases of haemophilia were reported from Rhodesia.[18,19] Also from East Africa, Lothe[20] diagnosed haemophilia in 7 Uganda Africans. All these reports suggest that haemophilia is not as rare in the Bantu as was initially suspected.

It is difficult to compare the incidence of disease in a developing population such as the Bantu with that in the developed populations of Europe. A significant proportion of the Bantu in the Transvaal live in remote areas, where the paucity of medical practitioners and the reluctance of primitive people to seek medical care in hospitals contribute to the apparent rarity of diseases such as haemophilia. Thus the incidence of haemophilia in the Bantu which might be computed from the present study is likely to be significantly less than the true incidence.

Further evidence that the actual incidence may be higher are the results of the factor VIII assays in this study. Thus, in 21 consecutive Bantu haemophiliacs where factor VIII was assayed, the level was less than 1% in 17 and 1 - 5% in 4. There were no patients with mild haemophilia (factor VIII 5 - 25%). This is in marked contrast to White populations, where in about 25% of haemophiliacs the disease is mild.[4] It would seem, then, that only the severe Bantu haemophiliac seeks medical aid. By the same token, the severe haemophiliac tends to present at an older age in Bantu than in Whites. Thus most severe haemophiliacs in Whites are diagnosed below the age of 5 years.[4] In the present series, however, many of the severe haemophiliacs were first seen in adolescence.

The ratio of haemophilia to Christmas disease was 8·4 : 1 in the present series. In the United Kingdom the ratio is 10 : 1, while in a review of the literature Jung[21] found a ratio of 4·5 : 1 over a total of 1,353 cases in White populations from a number of countries.

As in White haemophiliacs, haemarthrosis is the commonest presenting feature and the frequency of this complication similarly increases with increase of age. Gastro-intestinal and urinary tract haemorrhage is rare. There was not one instance of a gastro-intestinal or urinary tract bleed on presentation or on history. Cerebral haemorrhage did occur and was the cause of the single mortality in our records.

SUMMARY

Haemophilia has been regarded as rare in the Bantu. However, 67 cases were diagnosed over a 14-year period at the South African Institute for Medical Research, Johannesburg. The ratio of severe to mild forms was much higher in the Bantu compared with Whites, suggesting that the mild Bantu cases do not commonly seek medical help. It seems likely that haemophilia in the Bantu may be as frequent as in other ethnic groups.

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