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Idiopathic Thrombocytopaenia in Nigerian Children

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Summary

Lewis, M. J. and Essien, E. M. (1975). *Nigerian Journal of Paediatrics*, 2 (1), 9. **Idiopathic Thrombocytopaenia in Nigerian Children.** The clinical and haematological features of acute idiopathic thrombocytopaenic purpura (ITP), as well as the response to treatment in 13 children seen at University College Hospital, Ibadan, Nigeria over a five-year period are described. The general picture of the condition in the present series is one of an explosive illness, sometimes of great severity, but usually self-limiting and associated with a good prognosis.

IDIOPATHIC thrombocytopaenic purpura (ITP) is reported to be a rare disease among indigenous Africans and persons of African descent (Wintrobe, 1967; Jelliffe, 1970). The few reports of acute thrombocytopaenia in Africans concern *Onyalai* in East, Central and South Africa (Jelliffe, 1950; Laufer, 1953; Trowell, 1951; Wicks, 1972). Indirect references to ITP among Afro-Americans have been made by Newton and Zuelzer, (1951), and by Lusher and Zuelzer, (1966). This communication describes the features and management of the disorder in Nigerian children.

Materials and Methods

The series includes all the children with the diagnosis of ITP seen at the University College Hospital, Ibadan, over a period of five years (January, 1968 to December, 1972). Standard haematological methods were used. In each case the haematological diagnosis was made on the basis of a combination of the following criteria:

(*a*) Clinical evidence of haemostatic failure;

(*b*) A platelet count of less than 100,000/ul (Essien, Usanga and Ayeni, 1973);

(*c*) Bone marrow picture showing normal or increased number of megakaryocytes;

(*d*) Exclusion of known causes of thrombocytopaenia (drugs, aplastic anaemia, leukaemia and gross splenomegaly, etc.).

The clinical data included age, sex, month of presentation, symptoms and signs.

Results

The number of children with the diagnosis of ITP over the 5-year period was thirteen (6 male and 7 female). The age distribution (Fig. 1) shows that the youngest patient was 3 years old and the oldest 12 years. Ten of the patients were in the age group of 5–7 years. The month of presentation is shown in Figure 2. It will be observed that although there was no peak incidence in any one month, a majority of the cases presented between September and April.

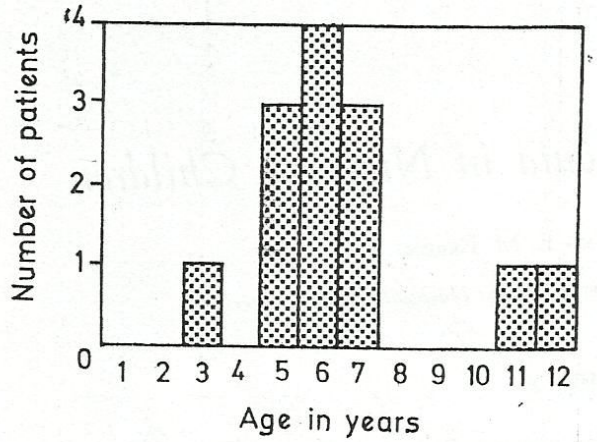

Fig. 1 Age distribution in 13 children with ITP.

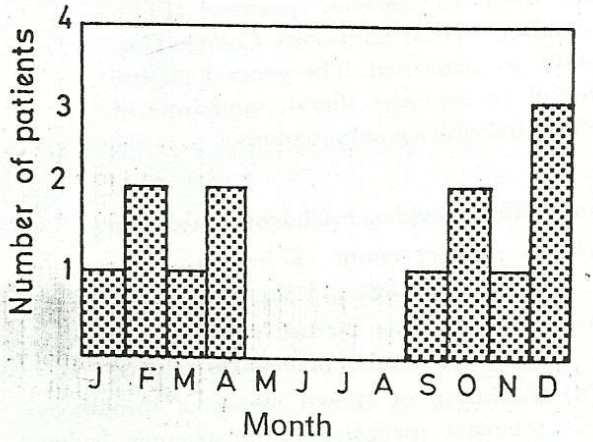

Fig. 2 Month of presentation showing that no patients were seen between May and August of each year. This suggests relative freedom during the wettest months of the rainy season.

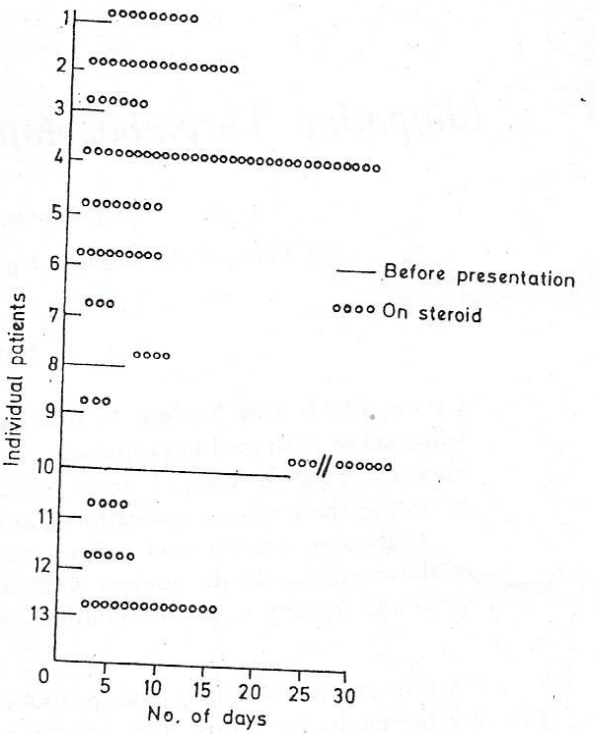

Fig. 3 Duration of illness before and after steroid therapy.
—— Duration of illness before presentation in hospital.
ooo Duration of steroid therapy before platelet count returned to normal or patient became lost to follow-up.

Clinical Features

Two patients gave a history of measles prior to the onset of bleeding. In the remaining eleven patients, there was no history of antecedent infections. The average period between the onset of symptoms and presentation at the hospital (Fig. 3) was five days (range 1–19 days). The sites of haemorrhage at the time of the first presentation are summarized in Table I. The commonest site of bleeding was in the mouth, particularly the gums; this occurred in 8 (62 percent) of the 13 patients. The next common site was the nose from where seven patients (54 percent) bled profusely. Cutaneous bleeding in the form of either purpura or ecchymoses occurred in 5 (38 percent) out of the 13 patients. Haemorrhagic bullae as described in *Onyalai* occurred in only one patient. There were 3 cases of haematuria. Miscellaneous bleeding sites included periorbital region, ulcer and abrasion in the leg, the lip, conjunctiva and traditional scarification sites.

Splenomegaly occurred in 5 patients (38 percent), with the spleen size varying between 1 cm and 4 cm below the costal margin.

Laboratory Findings

The main haematological findings are summarized in Table II. Thrombocytopaenia was present in all the 13 patients, and of these, 11

TABLE I

Sites of haemorrhage in 13 patients with ITP at time of presentation

Patient	*Skin*	*Nose*	*Mouth*	**Urinary Tract*	*Miscellaneous*
1	–	–	+	+	–
2	–	–	+	–	–
3	–	–	+	–	–
4	–	–	–	–	Periorbital
5	+	+	–	–	–
6	–	–	+	–	–
7	–	+	–	–	Leg Ulcer
8	–	+	+	–	Leg abrasion
9	–	+	+	–	–
10	+	–	–	–	Lip
11	+	+	+	+	Conjunctiva ethnic scarification
12	+	+	+	–	–
13	+	++	–	+	–

* Schistosoma haematobium was excluded as a possible cause of haematuria in all the patients.

TABLE II

Haematological Findings in ITP patients at first presentation

	WBC				*Bone Marrow*			
						Megakaryocytes		
Patient	*PCV %*	*Total /ul*	*Differential*	*Platelets /ul*	*Cellularity*	*Number*	*Platelet Production*	*Abnormal Cells*
1.	19	11,000	Normal	10,000	Not done	–	–	–
2.	23	12,400	Normal	10,000	Normal	Normal	Decreased	Eosinophilia
3.	11	7,100	Normal	88,000	Increased	Increased	Decreased	None
4.	46	Not done	–	10,000	Normal	Normal	Normal	None
5.	34	9,700	Normal	20,000	Normal	Normal	Normal	None
6.	25	Not done	–	10,000	Increased	Increased	Normal	None
7.	26	12,000	Normal	14,000	Increased	Increased	Normal	None
8.	23	7,100	Normal	34,000	Increased	Normal	Decreased	None
9.	17	9,400	Normal	12,000	Increased	Increased	Decreased	None
10.	11	19,000	Polymorph leucocytosis	12,000	Increased	Increased	Decreased	Megaloblastic
11.	32	9,500	Normal	18,000	Increased	Increased	Decreased	Megaloblastic
12.	29	10,400	Normal	10,000	Increased	Increased	Decreased	None
13.	22	12,000	Normal	15,000	Not done	–	–	–

patients had platelet count of 20,000/ul or less, Anaemia was present in the majority of the patients and the mean PCV was 24 percent. In ten patients, the PCV was below 30 percent. Five patients had leucocytosis (wbc > 11,000/ul).

Bone marrow examination was performed on eleven patients. Of these, seven had an increase of megakaryocytes with features of immaturity as described by Diggs and Hewlett (1948). In four patients, the megakaryocytes appeared quantitatively normal. Two of the bone marrow specimens showed early megaloblastic changes, and in one, there was slight eosinophilia. None of the bone marrow samples showed generalized hypoplasia.

Tests for platelet antibodies were not done.

Treatment

Nine patients required blood transfusion because of the severity of anaemia. All patients were routinely treated with prednisolone (2 mg/kg body weight/24 hours). There was clinical evidence of cessation of bleeding in all the patients within 1–4 days (mean 2 days) of starting steroid therapy. The average time between starting steroid therapy and achieving a normal platelet count in nine patients (Fig. 3) was 8.9 days (range 3–15 days). Two children did not gain normal platelet count and remained thrombocytopaenic 60 and 62 days respectively after starting on the prednisolone therapy. Both were lost to further follow-up. Only six patients attended for regular follow-up and in these, there was no recurrence of thrombocytopaenia for up to two years after initial presentation. Two of these six patients had splenomegaly initially, but this was no longer present six weeks later. Splenectomy was not performed in any of these patients.

Discussion

This report has revealed some striking clinical differences between the manifestations of acute idiopathic thrombocytopaenic purpura described here and those reported previously in other populations (Doan, Bouroncle and Wiseman, 1960; Lusher and Zuelzer, 1966). The first difference relates to the paucity of cutaneous manifestations. In this series, 38 percent of the patients presented with petechiae or ecchymoses. By contrast, in a study of three hundred and eighty-one patients reported by Doan, Bouroncle and Wiseman (1960), 70 percent presented with purpura in addition to bleeding from other sites. This difference may be due partly to the difficulty in recognizing tiny purpuric spots in a dark skin. Alternatively, the difference may reflect true paucity of cutaneous manifestations in ITP in the indigenous population. A possible explanation for this may be the relatively low normal platelet count in the local population as reported by Essien, Usanga and Ayeni, (1973). If this is the case, then the platelet count in indigenous patients with ITP have to be very low (<20,000/ul) before petechial haemorrhages occur. Further studies on this problem is in progress.

The second point of difference between ITP as described in this series and other published reports relates to the prevalence of splenomegaly which occurred in 38 percent of our patients. This is in marked contrast to a prevalence of only 2.6 percent in other series (Doan, Bouroncle and Wiseman, 1960). The size of the spleen in our series varied between 1 and 4 cm below the costal margin. Therefore, the presence of moderate splenomegaly in a patient with features suggestive of ITP is, in our view, not inconsistent with such a diagnosis in children who live in a holo-endemic malarious area.

It has been reported that acute infection with either plasmodium vivax or plasmodium falciparum may cause thrombocytopaenia, (Hill, Knight and Jeffercy, 1964; Skudowitz, *et al.*, 1973). In the present series, acute malaria infection would appear to be an uncommon cause of clinically apparent thrombocytopaenia since the incidence of thrombocytopaenia is

very low compared with that of malaria. None of our patients had laboratory evidence of malaria infection.

The third major difference is in relation to the high prevalence of anaemia which was severe (PCV less than 25 percent) in 54 percent of our patients, and moderate (PCV 25–30 percent) in an additional 22 percent of the cases. Thus in this series, severe anaemia was compatible with a diagnosis of ITP, whereas in other studies, (Baldini, 1966) the presence of severe anaemia was considered a contraindication to the diagnosis. There was no leucopaenia in any of the patients whose leucocyte count was performed. Four (30 percent) out of the thirteen patients had mild leucocytosis which may be a response to either the accompanying bleeding or an intercurrent infection. According to Wintrobe (1967) infection *per se* may cause anaemia. In our environment with widespread infections, it is possible that some of the patients in the present series had inapparent infection which together with the acute blood loss from severe thrombocytopaenia would lead to severe anaemia as occurred in a majority of our patients.

During the 5-year period of the present review, a total of 115,179 new patients attended the hospital. Of these 13 were cases of ITP, an incidence of 0.01 percent in the population. In a group of 132,235 patients reported by Doan, Bouroncle and Wiseman, (1960) the incidence of ITP was 0.18 percent. This difference is statistically significant ($P < 0.0001$), thus confirming the view that ITP is rare among indigenous Africans. The reason for this rarity is not clear.

The clinical course of the illness which was, in most cases, of short duration was consistent with the frequently transient nature of the disease in childhood (Karpatkin, 1971). All the patients were however, routinely placed on steroids in order to avoid possible intracerebral haemorrhage which has been reported to occur frequently in some Nigerian patients, (Essien and Adeloye, 1972). It is therefore, difficult to say how far the generally good response in our series was related to the steroids therapy. The clinical response following steroid therapy was, often dramatic, and in many instances antedated a rise in platelet count by several days. This is in accord with other published observations (Baldini, 1966). Two patients however, did not show this dramatic response; they remained thrombocytopaenic two months after the steroid therapy. Unfortunately, they were lost to follow-up. Although this initial poor response does not preclude eventual remission, perhaps these two patients were potential candidates for splenectomy.

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REFERENCES

Baldini, M. (1966). Idiopathic thrombocytopaenic purpura. *New Eng. J. Med.* **274,** 1245–1251.

Baldini, M. (1966). Idiopathic thrombocytopaenic purpura. *New Eng. J. Med.* **274,** 1360–1367.

Diggs, L. W. and Hewlett, J. S. (1948). A study of the bone marrow from thirty-six patients with idiopathic haemorrhagic (thrombocytopaenic) purpura. *Blood,* **3,** 1090–1104.

Doan, C. A., Bouroncle, B. A., and Wiseman, B. K. (1960). Idiopathic and secondary thrombocytopaenic purpura: clinical study and evaluation of 381 cases over a period of 28 years. *Ann. Intern. Med.* **53,** 861–876.

Essien, E. M. and Adeloye, A. (1972). Intracranial haemorrhage in haemophilia in Nigerians. *Trans. Roy. Soc. Trop. Med. Hyg.* **66,** 255–257.

Essien, E. M., Usanga, E. A., and Ayeni, O. (1973). The normal platelet count and platelet factor-3 availability in some Nigerian population groups. *Scand. J. Haemat.* **10,** 378–383.

Hill, G. J. II, Knight, V., and Jeffery, G. M. (1964). Thrombocytopaenia in vivax malaria. *Lancet.* **1** 240–241.

Jelliffe, D. B. (1950). Syndrome of Onyalai. *J. Trop. Med. Hyg.* **53,** 9–11.

Jelliffe, D. B. (1970). In Diseases of children in the Subtropics and Tropics, 2nd Edition, p. 547, Edward Arnold, London.

Karpatkin, S. (1971). Autoimmune thrombocytopaenic purpura. *Am. J. Med. Sci.* **261,** 127–138.
Laufer, W. E. (1953). Onyalai. *South Afric. Med. J.* **27,** 657–659.
Lusher, J. M. and Zuelzer, W. W. (1966). Idiopathic thrombocytopaenic purpura in childhood. *J. Pediat* **68,** 971–979.
Newton, W. A. J. and Zuelzer, W. W. (1951). Idiopathic thrombocytopaenic purpura in childhood, *New Eng. J. Med.* **245,** 879–885.
Skudowitz, R. B. Katz, J., Luric, A., Levin, J., and Metz J. (1973). Mechanisms of thrombocytopaenia in malignant tertian malaria. *Brit. Med. J.* **2,** 515
Trowell, H. C. (1957). Two cases of Onyalai in Uganda. *East Afr. Med. J.* **28,** 449–452.
Wicks, A. C. (1972). Onyalai—a disappearing disease entity. A case report and review of the literature. *Cent. Afr. J. Med.* **18,** 93–97.
Wintrobe, M. M. (1967). Clinical Haematology, 6th edition, p. 890. Henry Kimpton, London.