Hemoglobinopathies During Infancy in Western Orissa

G.C. Samal
J. Jena

Hemoglobinopathies, particularly sickle-cell anemia is regarded as a major cause of morbidity in pediatric practice in this part of Western Orissa(1,2). Other hemoglobinopathies encountered at times may mimic sickle-cell anemia and prove deceptive and refractory to the usual corrective measures. This may be particularly so during infancy as differentiation on usual methods are difficult due to high level of fetal hemoglobin and clinical features are at times inseparable. Hence, an attempt has been made to study the prevalance and distribution of various hemoglobinopathies as encountered during infancy in this part of Western Orissa.

Material and Methods

This study was conducted in the Department of Pediatrics, V.S.S. Medical College and Hospital, Burla, from January, 1977 to June, 1990. Five hundred anemic infants below one year of age and having hemoglobin below 10 g/dl were screened for the presence of hemoglobinopathies. A detailed recording of personal data, clinical history, physical examination and laboratory investigations were carried out as per a predesigned proforma. Investigations included were hematological indices, peripheral smear examinations, reticulocyte count, sickling test, fetal hemoglobin and hemoglobin electrophoresis(3). Serum bilirubin and radiological investigations were done whenever necessary.

From the Department of Pediatrics, V.S.S. Medical College, Burla, Sambalpur.

Reprint requests: Dr. G.C. Samal, 3R/49, Doctors Colony, Burla 768 017.

Received for publication October 23, 1990; Accepted April 18, 1991

Results

Of five hundred anemic infants, 32 showed abnormal hemoglobinopathies, the incidence being 6.4%. Different abnormal hemoglobinopathies seen were (*Table I*) sickle cell anemia (SS) in 12, sickle cell trait (AS) in 6 and 4 had sickle thalassemia (SF). Other hemoglobinopathies encountered were beta-thalassemia major (FF) and trait (AF) in 7 and 3 cases, respectively. Various clinical features encountered were depicted in *Table II*

Discussion

Though many surveys(2,4-6) have been carried out in different parts of the country to study the pattern of abnormal hemoglobinopathies, there are scarcely any reports during infancy. It is difficult to differentiate hemoglobinopathies during neonatal period because of high level of fetal hemoglobin. HbS is not normally demonstrable at birth since there is very little beta-chain synthesis by that time(7). In a

TABLE I—*Electrophoresis Pattern of Abnormal Hemoglobinopathies*

Age (mo)	SS	AS	SF	FF	AF	Total
0- 3	2	1	–	–	–	3
3- 6	2	2	1	4	–	9
6- 9	3	1	2	2	2	10
9-12	5	2	1	1	1	10
Total	12	6	4	7	3	32

TABLE II—*Clinical Features in Hemoglobinopatheis*

	Clinical findings	No	%
1.	Anemia	32	100.0
2.	Hepatomegaly	30	93.8
3.	Splenomegaly	28	87.5
4.	Fever	20	62.4
5.	Jaundice	15	46.9
6.	Bronchopneumonia	7	21.9
7.	Bleeding episodes	7	21.9
8.	Handfoot syndrome	5	15.1

previous study(2) out of the 17 'SS' infants, 4 were below three months of age. However, in the present study (*Table I*) out of the total 32 infants with abnormal hemoglobinopathies 12 (37.5%) had 'SS', 6 (18.75%) 'AS' and 4 (12.5%) had 'FS' pattern on electrophoresis. Of the 3 'SS' infants during first quarter of the year the youngest one was a seven days old male baby admitted with hyperbilirubinemia. Other hemoglobinopathies encountered were beta-thalassemia major in 7 (21.87%) and trait in 3 (9.38%) cases, respectively.

Clinical symptoms (*Table II*) common to both sickle-cell and thalassemia were anemia, hepatosplenomegaly, fever, and jaundice. Symptoms particular to sickle cell anemia were bleeding episodes and handfoot syndrome. Associated bronchopneumonic features were encountered in both varieties. Similar clinical features have been reported by others(2,4,8).

Examination of blood revealed marked anemia, normoblastemia and reticulocytosis with varying degrees of anisocytosis, poikylocytosis and polychromasia in all varieties of hemoglobinopathies. Target cells were particularly abundant in thalasssemia. Mean fetal hemoglobin and serum bilirubin were raised in both varieties. Similar blood pictures have been reported by others(2,4,8).

Although, detection and differentiation of hemoglobinopathies during infancy is a cumbersome process, efforts should be made to delineate the type of abnormal hemoglobinopathy so as to manage them effectively and adequately.

REFERENCES

1. Samal GC, Ahmed B, Behera SK. Incidence of sickle-cell disease. Pediatr Clin India 1978, 13: 33-36.
2. Samal GC. Sickle cell anemia during infancy. Indian Pediatr 1980, 17: 527-530.
3. Dacie JV, Lewis SM. Laboratory methods used in the investigations of hemolytic anemias. III. Hemoglobinopathies. *In:* Practical Hematology, 5th edn. Edinburgh, ELBS and Churchill Livingstone, 1975, pp 236-254.
4. Mitra SS. The clinical and hematological profile of thalassemia and hemoglobino-

pathies in India. Indian Pediatr 1983, 20: 701-703.

5. Bansal RK, Sharma B, Madhulika, Singh J, Saxena S. Hemoglobinopathies in anemic children of Western Rajasthan. Indian Pediatr 1983, 20: 1012-1015.
6. Magotra ML, Phadke MV. Anemias in infancy and childhood. Indian Pediatr 1975, 12: 493-498.
7. Willoughby MLN. Abnormalities of hemoglobin synthesis. *In:* Pediatric Hematology. Edinburgh, Churchill Livingstone 1977, pp 111-143.
8. Konotey Ahulu FID. The sickle cell disease: Clinical manifestations including sickle crisis. Arch Intern Med 1974, 133: 611-620.

Lymphangioma of Tongue

A.K. Basu
J. Basu
T.J. Banerjee
P.K. Sarkar

Lymphangioma of the tongue is an uncommon condition. Patients may present with either a cyst of the tongue or macroglossia. A large tongue causes difficulty in mastication and articulation. In newborn period an abnormally large tongue may cause serious feeding problems. We have observed three cases of lymphangioma of which two were cysts and one had macroglossia.

From the Departments of Pediatric Surgery and Anesthesiology, Medical College and Calcutta National Medical College, Calcutta.

Reprint requests: Dr. A.K. Basu, 90, Ballygunge Place, Calcutta 700 019.

Received for publication November 19, 1990; Accepted May 31, 1991

Case Reports

Case 1: A 3-day-old boy presented with a swelling protruding out of the mouth since birth. The swelling measured 7.5 by 5 cm and involved the anterior two-thirds of the tongue. It was cystic in consistency. Transillumination of the swelling was negative. There was no respiratory distress. Aspiration of the swelling showed turbid fluid. Microscopic examination of the fluid showed fat globules and pus cells; no organisms were cultured. After aspiration of 30 ml of fluid the swelling completely disappeared and the patient could close his mouth. He was treated with antibiotics and fed through Ryles tube. The swelling reappeared within a week but was smaller in size. The aspiration was repeated twice at four days interval; no further recurrence of the swelling was noticed.

Case 2: A 5-year-old girl presented with a gradually enlarged protuberent tongue since the age of 2 yr (*Fig. 1*). The patient could not close her mouth or speak coherently. The whole anterior two-thirds of the tongue was enlarged. The patient was treated by partial glossectomy. Under general anesthesia two overlapping hemostatic cum traction sutures of No/0 silk were placed at the root of the tongue and were tied tightly over two pieces of corrugated rubber drains, about two inches long to avoid cutting through (*Fig. 2*). The portion of the tongue to be removed was marked out on both the surfaces of the tongue with marking ink. A straight cutting needle was passed through the apex of the dorsal 'V' and was brought out through the apex of the ventral 'V'. The anterior two-thirds of the tongue thus marked out was removed using cutting diathermy. Hemostasis was secured after hemostatic sutures were removed one after the other. The remaining