ORIGINAL ARTICLE

The Pattern of Thalassemia in Children at the Department of Child Health, School of Medicine University of North Sumatera/ Dr. Pirngadi Hospital, Medan

by

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Abstract

A study of the characteristics of childhood thalassemia was conducted at the Sub Department of Pediatric Hematology, Dr. Pirngadi Hospital from June 1979 to May 1989.

There were 131 cases, consisting of 75 (57.25%) boys and 56 (42.75%) girls with an average of 12 admission every year. The predominant age group was 0–2 years, and the youngest was 3 months old. Javanese ethnic group appeared predominant in 36 (63.15%) cases.

Clinical symptoms of anemia were found in 112 (85.49%), hepatomegaly in 91 (69.46%), hepatosplenomegaly in 84 (64.12%), without enlargement of organ in 17 (12.97%), and icterus in 6 (4.58%). Hb-Electrophoresis was done in 42 cases, revealing 26 (61.90%) with thalassemia major, 15 (35.71%) Hb E thalassemia, and 1 (2.20%) Hb H thalassemia. Hb value at the first admission in 65 (49.62%) was less than 5 g/dl, in 63 (48.09%) it was 5-10 g/dl and in 3 (2.29%) more than 10 g/dl.

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Introduction

Thalassemia is a form of an inherited hemolytic anemia. It is inherited from both mother and father according to Mendel's law, and is caused by hemoglobin synthesis disorders (1,2). This disease is seen not only in the surrounding of the Mediterranian Sea but also in Asean countries, including Indonesia (3).

The clinical manifestations vary from the most severe thalassemia mayor to the asymptomatic thalassemia minor. In general, clinical manifestation of thalassemia mayor appears at the age of 6 months or more, where the child appears pale with abnormal physical development for age, hepatomegaly, bone disorders, and sometimes icterus. Thalassemia minor, in general, produces no marked clinical symptoms; it is sometimes accompanied by lymphatic enlargement (3,4). This study was designed to assess the characteristics of patients with childhood thalassemia admitted to the Sub Department of Pediatric Hematology, Department of Child Health, School of Medicine, University of North Sumatera/Dr. Pirngadi Hospital Medan. Dr. Pirngadi Hospital Medan.

Materials and methods

This study was done retrospectively by collecting and analyzing all medical records of patients with thalassemia admitted to the Sub Department of Pediatric Hematology, Department of Child Health, School of Medicine, University of North Sumatera/ Dr. Pirngadi Hospital Medan from June 1979 to May 1989.

The data collected consisted of age, sex, parents race, main complaints, laboratory findings, results of Hb-electrophoresis, bone marrow picture and body-weight.

The diagnosis was based on anamnesis (allo anamnesis), physical examination, peripheral blood, bone marrow and nutritional status of the patients. The nutritional status was determined by body weight for age.

For children under five years old the Kartu Menuju Sehat (Road to Health Chart) was used while for those more than 5 years old, the Harvard's International Standard (1959) was used. Patients with body weight of 80% of the 50th percentile were classified as normal (wellnourished), 60-80 % as mild and moderate and less than 60% as severe malnutrition.

Results

From June 1979 to May 1989, 131 patients (75 = 57.25%) boys and 56 = 42.75% girls) were admitted. Most of the cases were found in the age group of 0–2 years (47 cases = 35.87%) and it seemed that the older the child, the less was the number of cases found. The youngest patient was 3 months old and the oldest 14 years. Distribution of sex by the age group can be seen in Table 1.

The average of admission per years was 12 cases. The number of admission by sex is shown in Fig. 1.

Thirty six of 57 cases (63.15%) have a Javanese father as well as mother. Forty seven cases had the electrophoretic hemoglobin level showing 26 (61.91%), thalassemia mayor 15 (33.71%) Hb E thalassemia and 1 (2.2%) Hb-H thalassemia (Figure 2).

In 131 patients, 121 (85.49%) were admitted with symptomatic anemia, 107 (81.68%) with splenomegaly, 91 (69.47%) with hepatomegaly, 6 (4.58%) with icterus and 6 (4.58%) with the other complaints (Table 2).

Sixty five patients (49.62%) had Hb levels of less than 5 g/dl, 63 (48.09%) 5-10 g/dl, and only 3 (2.2%) more than 10 g/dl (Table 3).

Out of 131 patients, nutritional evaluation were only done in 123 patients revealing 21 (17.07%) wellnourished and 102 (82.93%) malnourished (moderate and poor) children (Table 4).

The average of admission per years was 12 cases. The member of admission by sex is shown in Figure 1.

Table 1 : *Distribution of patients by age and sex*

Age (years)	Sex		Number of patients
	Male	Female	
0 - 2	24	23	47
4	17	9	26
6	11	9	20
8	9	9	18
10	3	4	7
12	6	-	6
14	3	2	5
16	2	-	2
Total	75	56	131

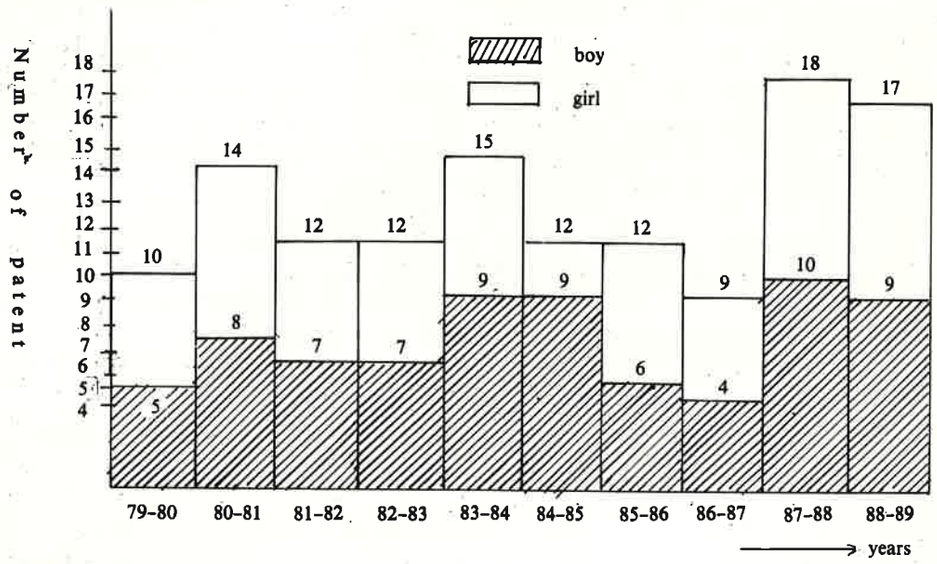

Figure 1 : *Number of patients per-year according to sex*

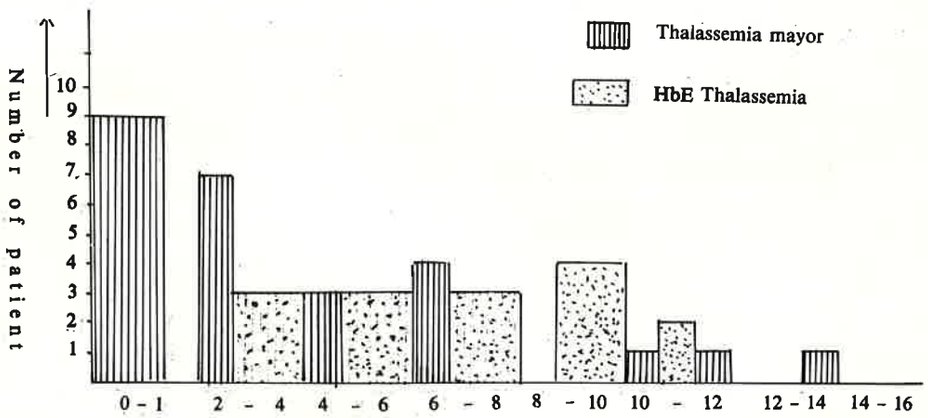

Figure 2 : *Number of patient with major thalassemia, Hb E thalassemia according to age group*

Table 2 : *Clinical manifestation*

Symptom / Sign	Number	%
Anemia	112	85.49
Splenomegaly	107	81.68
Hepatomegaly	91	69.47
Icterus	6	4.58
Others	6	4.58

Table 3 : *Hb Level*

Hb Level (g/dl)	Number	%
Less than 5	65	49.62
5 - 10	63	48.09
More than 10	3	2.2
T o t al	131	100.00

Table 4 : *Nutritional status by age group*

Age (years)	Nutritional Status						Number
	Good		Moderate		Poor		
0 - 1	8	34.78%	15	65.22%	0	0%	23
- 3	3	9.67%	19	61.29%	9	29.04%	31
- 5	6	27.27%	10	43.48%	7	20.25%	23
- 10	4	12.12%	23	69.69%	6	18.19%	33
10	0	0%	4	30.77%	9	69.23%	13
T o t a l	21	17.07%	71	57.72%	31	25.21%	123

Discussion

From June 1979 to May 1989, one hundred and thirty one patients were admitted (75 = 57.25% boys and 56 = 42.75%) girls). Iskandar (1979) found thalassemia in 58.2% boys and 41.8% girls.

Patients with thalassemia were predominantly found in the age group of 0–2 years (47 = 37.8%), this is similar with the report of some authors who stating that thalassemia occurred in the age group of 6 to 12 years at the time when the management of physiological anemia was unsuccessful.

In this study, the youngest patient was 3 months old when the electrophoretic Hb examination indicated the manifestation of thalassemia mayor. Iskandar also found the youngest patient to be 3 months old.

There was no increase in the number of admissions per year, with the average of 12 cases annually. Of 57 patients, it was found that 36 (63.15%) patients had a Javanese mother as well as father, while the rest were of various ethnic groups of North Sumatera. The predominance of the Javanese ethnic group needs further studies.

Electrophoretic Hb examination was done only in 42 cases showing 26 (61.90%), thalassemia major, 15 (35.71%) Hb E thalassemia and 1 (2.2%) Hb H thalassemia. It was found that thalassemia mayor occurred in the age group of 0–2 years (9 cases = 100%), and the number of patients with thalassemia mayor decreased with the increase of age. Hb E thalassemia occurred in the age group of older than two years. This might be caused by more severe course of thalassemia mayor Hb E thalasemia.

Anemia was found predominantly as a symptom at the first time of admission. This was found both in thalassemia mayor and thalassemia minor. In this study, 112 (83.49%) had anemia, 107 (81.68%) slenomegaly, 91 (69.47%) hepatomegaly, and 17 case showed no organomegaly at all.

The enlargement of organs is caused by the activities of extra medullary erythropoetic system and iron overloading of the organs. It might suppress the abdomen, especially the digestive tract; the child might lose its appetite and then it tends to the disturbances of nutritional status and growth.

In this study, only 21 (17.07%) were in good nutritional status, while 102 (82.93%) had malnutrition.

Conclusions

Of 131 patients with thalassemia in this study, 75 (57.25%) were boys and 56 (42.75%) girls. Most of these patients were found in the age group of 0–2 years. The youngest patients was 3 months old. Most of the cases are of the Javanese ethnic group. The symptoms of thalassemia mayor might occurr in the group of younger than 2 years. The main clinical manifestation in these cases were anemia, splenomegaly and hepatomegaly. Patients might have no enlargement of organs. Generally, patients were admitted with malnutrition.

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