Prevalence of thalassaemia in schoolchildren in north-eastern Badia, Jordan

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SUMMARY The prevalence of the different types of thalassaemia and that of iron deficiency anaemia was investigated in 1020 schoolchildren (age range: 6–15 years) from the north-eastern Badia region of Jordan. β-thalassaemia minor was the most prevalent (3.04%). followed by α_2-thalassaemia (2.06%). Only three cases of α_1-thalassaemia and one case of β-thalassaemia major were found. Iron deficiency anaemia was diagnosed in 54 children (5.3%) (33 males, 21 females). The mean values of the blood characteristics of the normal, haemoglobinopathic and iron-deficient children were examined and compared.

Introduction

Haemoglobinopathies, disorders of haemoglobin, are the most common single-gene disorders in the world's population [*1,2*]. Haemoglobinopathies can be divided into two main groups: structural variants (e.g. sickle-cell haemoglobin) and disorders of synthesis (e.g. thalassaemia)

Thalassaemia can be classified according to the reduction of one or more of the globin chains of haemoglobin into two major types: α-thalassaemia and β-thalassaemia [*3*]. Heterozygous β-thalassaemia and both α_1- and α_2-thalassaemia are asymptomatic and can only be discovered incidentally in the course of family studies or population surveys [*4*]. During the past 25 years, surveys for haemoglobinopathies have been conducted in most Arab countries. Many of these studies have documented relatively high frequencies of β-thalassaemia (4.6% in Libyan Arab Jamahiriya, 4.49% in Tunisia, 1%–3% in Lebanon, Algeria and Morocco, and 3.0% among ethnic Saudi Arabs) [*4,5*]. Although the incidence of α-thalassaemia in the populations of Mediterranean Arabic-speaking countries has not been fully evaluated, relatively high rates of incidence have been reported in Tunisia (4.8%) and Algeria (9%) [*5*]. Heterozygous α_2-thalassaemia and homozygous α_2-thalassaemia have been found in north-western Saudi Arabia at frequencies of 0.121% and 0.0046% respectively [*6*].

In Jordan, the only evaluation of the incidence of haemoglobinopathies was that of Bashir et al., who estimated the prevalence of β-thalassaemia and α-thalassaemia in the northern Jordan valley at 3.3% and 3.5% respectively [*7,8*].

The aim of our study was to survey the incidence of haemoglobinopathies in

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schoolchildren aged between 6 years and 15 years in the north-eastern Badia region of Jordan.

Subjects and methods

During the course of this investigation, blood samples were collected from 1020 healthy male and female children from 29 schools serving 32 villages between the towns of Al-Mafraq and Al-Safawi, in north-eastern Badia, Jordan. As the schools are segregated, female and male schools of approximately equal numbers of students were selected for investigation. Blood samples were randomly collected from between 5 and 10 non-related pupils chosen from each class in grades 1–9.

The blood samples were collected in EDTA vacutainer tubes. A Coulter Counter T-660 was used to determine the main characteristics of the blood: mean corpuscular volume (MCV); haemoglobin concentration (Hb); packed-cell volume (PCV); red blood corpuscles (RBC); white blood cells (WBC) and blood platelets (Plt). Samples where the MCV was less than 80.4 fL were subjected to electrophoretic separation of haemoglobin on cellulose acetate paper [*1*]. Microcolumn chromatography was used to separate the Hb A_2 [*9*], and the presence of Hb H was detected by cresyl blue staining [*10*]. α_1-thalassaemia was indicated by the presence of Hb H bands and confirmed by the appearance of "golf-cells" following staining with brilliant cresyl blue. Otherwise, these cases exhibited normal blood characteristics.

β-thalassaemia minor trait was diagnosed in cases with abnormal RBC and elevated Hb A_2 (> 3.5%) [*1*]. Calculation of the discriminant factor from RBC indices, according to England–Frazer [*11*], was used to distinguish β-thalassaemia minor (negative values) from iron deficiency anaemia [*1*]. β-thalassaemia major was diagnosed according to the appearance of

Table 1 **Mean values of blood characteristics of the normal subjects in north-eastern Badia, Jordan, 1997**

Parameter	Mean ± *s* Males (*n* = 389)	Females (*n* = 269)	Total (*n* = 658)
Hb (g/dL)	13.42 ± 0.84	13.36 ± 0.77	13.40 ± 0.81
Packed cell volume (L/L)	0.41 ± 0.02	0.40 ± 0.02	0.41 ± 0.02
Mean corpuscular volume (fL)	84.08 ± 2.59	84.83 ± 3.17	84.39 ± 2.86
Mean corpuscular Hb (pg)	27.73 ± 1.10	28.16 ± 1.23	27.91 ± 1.17
Mean corpuscular Hb concentration (g/dL)	32.99 ± 0.74	33.18 ± 0.76	33.06 ± 0.75
Red blood cell count (× 10^{12}/L)	4.84 ± 0.30	4.75 ± 0.29	4.80 ± 0.30
Platlets (× 10^9/L)	336.32 ± 74.63	331.15 ± 69.43	334.20 ± 72.54
White blood cell count (× 10^9/L)	8.71 ± 2.23	8.86 ± 2.38	8.77 ± 2.29

s = *standard deviation*
Hb = haemoglobin

Hb F electrophoretic bands. Its relative abundance was determined by alkaline denaturation [*10*]. Serum iron concentration and total iron-binding capacity (TIBC) were measured in the samples indicated as possible cases of iron deficiency anaemia [*11*].

Results were analysed using *SPSS*.

Results

In 362 of the 1020 children (35%), MCV was less than 80.4 fL, and 110 of these children (10.8%) exhibited different haemoglobinopathies. Of the 110 haemoglobinopathic children, 56 were diagnosed as having α- and β-thalassaemia, and 54 suffered from iron deficiency anaemia. RBC indices for the 110 haemoglobinopathic children were significantly different from those of normal children (n = 658) ($P < 0.001$, Student t-test).

The haematological characteristics of the study population are shown in Table 1. Table 2 shows the characteristics of the different haemoglobinopathies. In 250 children with MCV less than 80.4 fL, mean values of the haematological characteristics were within normal range and there was no evidence of haemoglobinopathic disorders. β-thalassaemia minor was the most prevalent of the haemoglobinopathies (diagnosed in 3.04%). α_2-thalassaemia was moderately prevalent (2.06%), while α_1-thalassaemia was diagnosed in only three children where Hb H bands were evident following electrophoretic separation of haemoglobin. Two of these children were males. Blood samples from their fathers also showed Hb H bands. The third case of α_1-thalassaemia was a female

Table 2 **Haematological characteristics associated with thalassaemia conditions in schoolchildren in north-eastern Badia, Jordan, 1997**

Parameter	α_1-thalassaemia		α_2-thalassaemia		β-thalassaemia	
	Males (n = 2)	Females (n = 1)	Males (n = 18)	Females (n = 3)	Males (n = 16)	Females (n = 15)
Hb (g/dL)	12.10±0.01	12.70	13.25±1.13	13.77± 0.40	12.16±1.49	12.11±1.46
Packed cell volume (L/L)	0.37±0.01	0.38	0.41±0.02	0.42±0.02	0.39±0.04	0.38±0.03
Hb A_2 (%)	3.24±0.01	2.01	2.53±0.46	2.82± 0.42	4.75±0.89	4.11±0.61
Mean corpuscular volume (fL)	80.40±0.05	80.30	72.66±4.83	75.17± 1.22	67.56±7.36	69.18±7.07
Mean corpuscular Hb (pg)	26.61±0.91	26.08	23.44±2.53	24.69± 0.86	21.38±3.14	21.83±3.06
Mean corpuscular Hb concentration (g/dL)	3.11±0.58	33.30	32.04±1.46	32.88± 0.67	31.56±1.29	31.42±1.28
Red blood cell count (× 10^{12}/L)	4.55±0.16	4.87	5.67±0.22	5.58± 0.34	5.74±0.52	5.58±0.45

Values are expressed as mean ± standard deviation.
Hb = haemoglobin

Table 3 **Means and ranges of values of some blood characteristics and iron levels associated with iron deficiency anaemia in schoolchildren in north-eastern Badia, Jordan, 1997**

Parameter	Mean ± *s*			Range
	Males (*n* = 33)	Females (*n* = 21)	Total (*n* = 54)	
Hb (g/dL)	11.42 ± 0.30	10.79 ± 1.72	11.18 ± 1.40	5.80 – 12.80
Packed cell volume (L/L)	0.36 ± 0.03	0.34 ± 0.04	0.36 ± 0.03	0.22 – 0.40
Hb A_2 (%)	2.68 ± 0.53	2.45 ± 0.64	2.57 ± 0.59	0.86 – 3.40
Mean corpuscular volume (fL)	72.99 ± 3.90	70.44 ± 6.45	72.00 ± 5.15	56.00 – 76.40
Mean corpuscular Hb (pg)	23.19 ± 1.98	22.19 ± 3.00	22.80 ± 2.45	15.06 – 25.94
Mean corpuscular Hb concentration (g/dL)	31.71 ± 1.32	31.38 ± 1.69	31.58 ± 1.47	26.36 – 34.35
Red blood cell count ($\times 10^{12}$/L)	4.94 ± 0.30	4.86 ± 0.37	4.91 ± 0.33	3.85 – 5.64
Iron (µg/dL)	50.23 ± 14.77	40.89 ± 14.64	45.92 ± 15.17	21.00 – 75.10
TIBC (µg/dL)[a]	377.82 ± 66.05	438.11 ± 85.87	405.65 ± 80.33	260.00 – 646.80
Transferrin saturation (%)[a]	13.51 ± 3.79	9.65 ± 4.08	11.73 ± 4.32	4.56 – 16.50

[a] *Values for males and females were significantly different (P < 0.05) (Student* t*-test).*
s = *standard deviation* Hb = *haemoglobin*
TIBC = total iron-binding capacity

whose mother was also α_1-thalassaemic. Apart from the single female diagnosed with α_1-thalassaemia, the cases of α_2-thalassaemia showed the lowest levels of Hb A_2 among the haemoglobinopathic children (Table 2).

No differences attributable to gender were observed in the relative prevalence of β-thalassaemia minor. However, only 3 of the 21 children diagnosed with α_2-thalassaemia were female. In cases of either haemoglobinopathy, the MCV was slightly higher in females compared with males, but the differences between mean MCV values for the two sexes were not statistically significant ($P > 0.05$) (Table 2).

It is noteworthy that β-thalassaemia major was found only in one 10-year-old boy during this study. Electrophoretic separation of a blood sample from the affected boy revealed the presence of fetal haemoglobin, which according to the alkaline denaturation method [*1*] amounted to 2.8% of the total haemoglobin. The red corpuscles of this β-thalassemic major child were normoblastic, anisopoikilocytotic and hypochromic with numerous target cells.

Iron deficiency anaemia was diagnosed in 33 males and 21 females (5.3% of the total). In these cases, MCV varied from 56.0 fL to 76.4 fL with a mean value of 72.00 ± 5.15 fL. Haemoglobin concentration and RBC count were, expectedly, lower than normal (Table 3). The level of Hb A_2 ranged from 0.86% to 3.40% in cases of iron deficiency anaemia (mean value 2.57 ± 0.59%), comparable to that observed in cases of α_2-thalassaemia (Tables 2 and 3). Although the

number of males diagnosed with iron deficiency anaemia was greater than females, no statistically significant differences between the sexes were observed for any of the blood characteristics examined except for TIBC and transferrin saturation ($P < 0.05$, Student t-test). Table 3 shows these results.

Discussion

The mean values of blood characteristics of the study population were generally comparable to the normal values for other populations. There were few intraspecific differences except in the cases diagnosed with haemoglobinopathies. The prevalence of β-thalassaemia minor indicated in the present study (3.04%) is comparable to that reported for the population of the Jordan Valley (3.1%) [7], northern Jordan (3.5%) [7,8] and Saudi Arabia (3.0%) [5]. In Lebanon, another neighbouring country, the reported prevalence of β-thalassaemia minor (1.7%) is much lower [5]. The incidence of β-thalassaemia major is apparently very low. Only one case was found during our study, and no record of incidence could be traced for this area. The incidence of α-thalassaemia among the Bedouin of north-eastern Badia (2.3%) was lower than that reported for northern Jordan [7]. Three cases with typical α_1-thalassaemia characteristics were diagnosed, indicating a quite low level of prevalence of this type of thalassaemia in the area of study, compared with other parts of the Eastern Mediterranean Region. Higher rates have been reported from other Mediterranean countries (e.g. 7%–8% in Greece) [12]. Apart from a few studies in Israel, which investigated the genetic and ethnic implications of these disorders [13], almost all other studies in the region have been limited to estimations only of the prevalence of these diseases [14].

The prevalence of iron deficiency anaemia in this area has not received much attention and no studies of its occurrence in schoolchildren could be traced. However, according to Amine and Al-Awadi [15], the prevalence of anaemia in preschool children has declined in the last 2 decades, although a good proportion of the children are still anaemic. These authors observed a prevalence of dietary anaemia of about 30% in Kuwait. In our study, a prevalence of only 5.4% was observed for iron deficiency anaemia among schoolchildren in north-eastern Badia. A higher prevalence (11.2%) has been observed among schoolchildren in neighbouring Syrian Arab Republic [16].

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