

Growth assessment in thalassaemic patients in Mosul

Nahlah Wadullah Hasan*, Alaa Abd-Al-Ghany Younis and Enaam Khazal Shihab

Doctor specialist / Diploma/ community medicine Directorate of Health / Nineveh, Kwestan, Erbil, Iraq

*Corresponding author:

Nahlah Wadullah Hasan

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Abstracts: Thalassaemia syndrome is a series of genetic disorders of haemoglobin synthesis. It is one of the major public health problems. The study is aiming for assessing the thalassaemia effect on growth. A case control study was implemented in thalassaemia center at Ibn Al-Atheer hospital during the period from 2000 Dec-2001 Jan where 202 cases of the attendants have been involved in the study and equal number of children attending to the outpatient for other causes as a control. Male consist 60.8% of the cases and 49.2% of the control while the female where 39.3% of the cases and 50.8% of the control. 43.6% of the cases fall in the age group >5-10 years and the highest percentage of the control 48.2% in the same age group.

Keywords: Thalassaemia syndrome, Ibn Al-Atheer hospital, Mosul.

INTRODUCTION

Thalassaemia syndrome is a series of genetic disorders of haemoglobin synthesis which is inherited on autosomal recessive basis

There are two types of thalassaemia:

1. alpha thalassaemia
2. Beta thalassaemia which is subdivided into three types :
 - a-thalassaemia major
 - b-thalassaemia intermediate
 - c- thalassaemia minor .

Epidemiology

Thalassaemia is widely distributed throughout the world and is one of the major public health problems. Thalassaemia primarily affects people of Mediterranean, Africa and Asia. Survey in Iraq showed that beta thalassaemia trait is carried by 4.5 – 5 % of the population, in Nineveh province more than patients were registered in thalassaemia center since 1997.

Beta thalassaemia major:

Beta thalassaemia is the most important one and is characterized by progressive anemia which is

manifested during the second six months of life associated with marked splenomegaly and feature of chronic hemolytic anemia, paler, jaundice, the spleen and liver are enlarged by extra medullary hematopoiesis , the bone marrow become thin and pathological fracture may occur, massive expansion of bone marrow of the face and skull produced characteristic mongoloid facies. The growth is impaired in older children, puberty delayed or absent because of secondary endocrine abnormality also diabetic mellitus and cardiac complication have been common terminal event. Treatment of this disorder depends mainly on adequate blood transfusion, desferrioxamine, folic acid, vitamin C, splenectomy and bone marrow transplantation, but the disease remain incurable.

Beta Thalassaemia Intermedia:

Thalassaemia intermedia means mild type of the disorder in which the patient can survive without regular blood transfusion the patients are either completely healthy or have mild anemia which may become severe after two years of age.

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Beta Thalassemia Minor:

It means the heterozygous state of thalassemia in which the patients show no symptoms and is diagnosed accidentally during medical examination for unrelated symptoms . these patients may develop symptoms during period of infection or pregnancy .

Skeletal lesion in beta thalassemia major : untreated or blood transfused children with beta thalassemia major show skeletal lesions due to hyper expansion of bone marrow this causes shortening of the upper arms and premature fusion of epiphyseal line , spinal deformities with kyphosis, scoliosis and vertebral collapse . while in treated patients with high transfusion program and iron chelation drug has drastically improved their life expectancy and quality of life and reduced the incidence and severity of skeletal lesions and other complications.

Cause of Growth Retardation in Beta Thalassemia:

Despite regular transfusion and desferrioxamine treatment for thalassemic patients , growth failure is commonly seen during adolescence due to reduce in growth hormone secretion and insulin like growth factor (IGF-1) in response to neurosecretory dysfunction due to iron overload some thalassemia patients. Another cause of growth retardation in beta thalassemia major is chronic anemia , iron overload, desferrioxamine toxicity , malnutrition and emotional caused .

Aim of study

General aim: assessment of the thalassemia effect on growth.

Specific Aim:

1. To determine the age and sex distribution of the cases and control
2. To measure the progress of growth by height , weight / age of the studied cases between the year 1998 and the year 2000 .
3. To compare the height, weight /age for the cases and control for the year 2000.
4. To find the relation between thalassemia major and the consanguinity of the parents .
5. Recommending program for prevention and control of the disease.

MATERIAL AND METHOD

A case control study was implemented in thalassemia center at Ibn Al-Atheer hospital during the period from 2000Dec-2001Jan where 202 cases of the attendants have been involved in the study and equal number of children attending to the outpatient for other causes as a control .

STUDY DESIGN

The Study Composed Of Two Parts:

- 1- comparing the growth of patient for the last three years (1998-2000) by finding the height and weight of the patient.
- 2- comparing the growth of the patient for the year 2000 with the control group for the same year

DATA SOURCE:

A-questionnaire form was prepared which included information (age , sex , age of diagnosis ,consanguinity of parents ,body height , body weight) by interviewing parents or cases who were attending thalassemia center B – recorded information of the year 1998 which is taken from the recorder for the same sample cases (age , sex , age of diagnosis , height/age , weight/age).

Data Analysis

Data analysis has been done by tabulation of data and statistical test

- Chi square X2.
- Z-test two proportion.
- Odd’s ratio.

RESULTS

The result of this study which was carried out in thalassemia center. 202 cases were assessed for their growth for the last three year 1998-2000 and the same number attended at an outpatient 202 was taken .the result of study sample the cases and control were found that male consist 60.8% of the cases and 49.2% of the control while the female where 39.3% of the cases and 50.8% of the control . 43.6% of the cases fall in the age group >5-10yrs and the highest percentage of the control 48.2% in the same age group.

and age among 79 case and 103 control as expresses by p-value and odd’s ratio :

height percentile	>3-5		5-10		10-15		15-20		Total no.		P-value	Odd’s ratio
	Case %	Cont. %	Case%	Cont. %	Case %	Cont.%	Case%	Cont. %	Case %	Cont. %		
<3	6.3	30.7	36.1	30.7	46.8	38.4	10.6	----	47	13	<0.001 v.h.s	11.5
3-10	-----	30	30.7	40	38.4	30	30.7	-----	13	20	NS	1.5
10-25	20	5.5	70	66.6	10	22.2	----	5.5	10	18	NS	0.5
25-50	40	22.7	60	50	----	22.7	--	4.5	5	22	0.001	4.01
50+	25	13.3	75	46.6	-----	20	--	20	4	30	<0.001	7.7

Z-test two proportion

Table 2: the association between male height and age among 123 case and 100 control as expresses by p-value and odd's ratio:

Height percentile	>3-5		5-10		10-15		15-20		Total no.		P-value	Odd's ratio
	Case %	Cont. %	Case%	Cont. %	Case%	Cont.%	Case%	Cont. %	Case%	Cont. %		
<3	7.8	30.7	34.3	23	39	38.4	18.7	7.6	64	13	<0.001	7.25
3-10	14.2	41.6	61.9	16.6	23.8	25	--	16.6	21	12	NS	0.6
10-25	25	14.4	41.6	57.1	29.1	23.8	4.1	4.7	24	21	NS	0.9
25-50	36.3	13.6	45.4	50	18.1	22.7	--	13.6	11	22	0.001	2.8
50 ⁺	33.3	18.7	66.6	65.6	--	12.5	--	3.1	3	32	<0.001	18.8

Z-test two proportion

Table 3: the association between female weight and age among 79 case and 103 control as expresses by p-value and odd's ratio

Weight percentile	>3-5		5-10		10-15		15-20		Total no.		P-value	Odd's ratio
	Case%	Cont.%	Case%	Cont.%	Case%	Cont.%	Case%	Cont.%	Case%	Cont.%		
<3	6.8	11.7	25	47	50	38.2	18.1	2.9	44	34	<0.001	2.5
3-10	11.1	30.7	66.6	53.8	16.6	15.3	5.5	--	18	13	NS	2.04
10-25	12.5	22.2	50	62.9	73.51	14.81	--	--	8	27	0.001	0.3
25-50	28.5	21.4	71.4	28.5	--	42.8	--	7.1	7	41	NS	0.6
50 ⁺	--	20	100	33.3	--	6.7	--	40	2	15	<0.001	6.5

Z-test two proportion

Table4: the association between male weight and age among 123 case and 100 control as expresses by p-value and odd's ratio

Weight percentile	>3-5		5-10		10-15		15-20		Total no.		P-value	Odd's ratio
	Case%	Cont.%	Case%	Cont.%	Case%	Cont.%	Case%	Cont.%	Case%	Cont.%		
<3	10.4	13.3	25	60	39.5	20	25	6.6	48	15	<0.001	3.6
3-10	20.8	31.8	41.6	40.9	37.5	22.7	--	4.5	24	22	NS	0.8
10-25	16.6	32	50	44	29.1	20	4.1	4	24	25	NS	0.72
25-50	5.8	5.8	76.4	58.8	17.6	29.4	--	5.8	17	17	NS	0.78
50 ⁺	40	14.2	50	47.6	10	19	--	19	10	21	0.001	3

Z-test two proportion

Table 5: relationship between consanguinity and age of diagnosis of 202thalassemic patients .

Age of diagnosis (years)	Positive relation	Negative relation	Total
0.5-1	117 (73.5)	42 (26.6)	159 100
>1->2	29 (67.4)	14 (32.6)	43 100
Total	146	56	202

DISCUSSION

The sample of the study was taken from attendants by systemic random sampling the control were taken from attendance of outpatient of the same hospital from which the cases were selected weight , height measurement were compared for the sample cases in the year 1998 and 2000 to see the progress during these three years then the cases were compared with the control to measure the difference between normal growth and thalassemic patient. The mean age of the cases and the control (9.6,8.5yrs) were nearly equal and the children below 10yrs constitute 56.3% ,68.6% for the case and the control respectively probably because most of the patients die after the end of the first decade of life . the cause of death is mostly due to anemia , infection , and complication of hypersplenism.

In comparing weight / age percentile for male and female for the percentage of the year 1998 and the year 2000 marked improvement were observed which is probably due to good quality care in the newly

established thalassemic center in Mosul with strict follow-up with patients.

Growth retardation was evaluated according to the growth and development chart whether they are above or below third percentile in other word 3% below the lower limit was regarded as exceptional and therefore not included in the normal range.

The stunting growth as the child become older for both male and femal cases which is found in our study is agreement with the study in Ibn-Al- Balady Hospital.

CONCLUSION

The disease should be prevented rather than treated because even in a good center the treatment remain symptomatic and the patients growth remains retarded and handicapped the following point were concluded:

1. The number of cases was higher in male than female.
2. The majority of our patients were growth retarded . retardation of height more than that for weight and growth retardation was more evident in female than in male .
3. Consanguinity is more prevalent in our patient specially first cousin marriages were most of the cases are diagnosed in the first year of life.

RECOMMENDATIONS

A-Through primary health care centers (PHC)

- 1- Primary prevention by health education for community and family through Audio-visual education facilities, and making small council circular showing the danger of the disease and to be given to the concerned and future parents
- 2- Population screening for heterozygous beta thalassemia for the assessment of HbA2 level

B- Through family planning centers:

- 1-Obligatory pre-marital test for the assessment of carrier , clearly the carrier must be warned about the danger of marriage to a similarly affected individuals.no affirm of wedding should be done without medical certificate.
- 2 –Advice to the parent who have already had one abnormal child to use contraceptive.

C- Through thalassaemic center:

- 1- The growth of thalassaemic patient should be monitored routinely at every follow up visit and documented on growth chart to detect early changes

- 2- Establishment of an appropriate protocol for investigation and treatment for all thalassaemic patient

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