Correlation of Wrist Circumference as a Sign of Severity with Significant Splenomegaly in Patients with Beta Thalassemia Major

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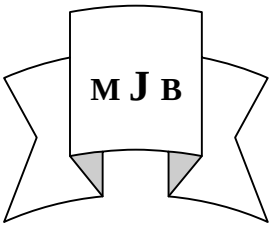

Abstract

Objective: Bone changes are relatively common in improperly treated β-thalassemia major. One of the signs of severe thalassemia is significant splenomegaly.The aim of the study is to see the correlation of wrist circumference with splenomegaly as a sign of severity.

Methods: One hundred and five patients with thalassemia major were studied,52 patients with significant splenomegaly and 53 without splenomegaly.Wrist circumference was measured for both groups. Fifty nine healthy children were taken as a control regarding wrist circumference.

Results: One hundred and five patients with thalassemia were studied. Growth parameters of the group with splenomegaly were found below the standard for their age and sex and wrist circumference was found to correlate significantly with splenomegaly as a sign of severity($P<0.05$) and the confidence interval was 95%.

Conclusion: Wrist circumference is a measurable, reliable index to assess severity of thalassemia major.

مقارنة محيط الرسغ بتضخم الطحال كعلامة شديدة في فقر دم البحر الأبيض المتوسط

الخلاصة

تعد التغيرات التي تحصل في عظام المرضى المصابين بفقر دم البحرالابيض المتوسط (الثلاسيميا) شائعة و يعتبر تضخم الطحال من العلامات التي تدل على شدة المرض.

الهدف من الدراسة :هو مقارنة محيط الرسغ مع تضخم الطحال كعلامة شديدة في مرضى الثلاسيميا.

تم اخذ عينة من 105 مريضا مصابا بالثلاسيميا الشديدة عند مراجعتهم لمركزالثلاسيميا في مستشفى بابل التعليمي للنسائية و الأطفال و قسمت الى 52 مريضا مصابا بتضخم الطحال و قورنت مع 53 مريضا مصابا بنفس المرض مع عدم وجود لتضخم الطحال كما تم اخذ عينة أخرى لأطفال أصحاء لإبعاد عامل تغير محيط الرسغ بسبب تغير العمر.

تم قياس محيط الرسغ لليد اليمنى كقياس ثابت من قبل طبيب أطفال اختصاص وقورنت النتائج للقراءات الثلاث و كانت نتائج المقارنة معنوية$P<0.05$

الاستنتاج:

محيط الرسغ قياس سهل قابل للتطبيق و من الممكن الاعتماد عليه لمعرفة شدة مرض الثلاسيميا اذا ما قورن بتضخم الطحال كعلامة شديدة.

Introduction

Thalassemia covers a group of hemolytic anemia caused by inheritance of abnormality of hemoglobin production. Beta thalassemia major occurs when the patient is homozygous for abnormal hemoglobin synthesis associated with defective Beta chain production and persistent fetal hemoglobin synthesis.

Beta Thalassemia major is characterized by ineffective erythropoiesis leading to severe anemia and bone marrow expansion which gives thalassemic patients their characteristic faces and bone widening[1]. Treatment with blood transfusion and chelation therapy have made patients survive longer[2], but it has left them with the bad effects of iron deposition in skeleton as it has been assessed by MRI [3] and it's deposition in endocrine organs[4], with the result of poor growth and sexual development [5,6], however hormonal treatment has improved bone density[7]. Fractures in the vertebrae and long bones may occur due to osteoporosis[8]which might affect up to a half of patients with thalassemia major[9] .Sclerotic changes of long bones and hyper-reactivity of the bone marrow have been described with desferoxamine therapy[10-12]. Splenic enlargement of 6 cm or more is nearly always accompanied by some hypersplenism [13] and a spleen larger than this should be removed[14].Skeletal changes have been reported in patients with thalassemia major, it was attributed to bone marrow hyperplasia and widening of the marrow space[15,16]. Wrist bones are part of the skeleton and they may be affected by changes like any bone in the body not only in thalassemia but it has been described in rickets and achondroplasia,but wrist size in thalassemia has not been studied as a sign of progressive disease as it is with the case of splenomegaly .

The aim of the study is to use the wrist circumference as a sign of severity if compared to splenomegaly which is by it self an indicator of severity.

Patients and Methods

Hundred and five patients with thalassemia major attending thalassemia centre at Babylon Maternity and Children Teaching Hospital for blood transfusion were studied in the period June&Jully 2008, age:2-14 years. They were 52 patients with significant splenomegaly (more than 6 cm below the costal margin), 58 patients with mild or no splenomegaly and 59 healthy children taken as control for normal wrist circumference Patients criteria were reviewed during their visit to the centre including their Hb-electrophoresis,blood requirement /year, splenectomy and growth parameters. Wrist circumference of the right arm of both groups was measured at the widest bony prominence by a consultant pediatrician interested in thalassemia and a control group of 59 healthy children was taken for comparison .
Statitical analysis was based on analysis of variance (ANOVA) and LSD pair wise comparison[17].

Results

There were one hundred and five patients with thalassemia major

attending the centre for follow-up and blood transfusion. Fifty two patients were found to have significant splenomegaly or they have their spleen removed and fifty eight patients were found to have mild or no splenomegaly. The characteristics of thalassemic patients with splenomegaly are presented in table(1), their hieght is obviously delayed which indicates severity of the disease if linked to splenomegaly.

Table 1 Patients criteria

Criteria	No	%
Sex		
Male	35	(67.3%)
Female	17	(32.7%)
Age in years		
2-4	4	(7.7%)
5-7	13	(25%)
8-10	15	(28.8%)
11-13	13	(25%)
>13	7	(13.5%)
Height of patients with splenomegaly		
<3rd centile	35	(67.3%
<50th centile	10	(19.2%)
<90th centile	7	(13.5%)

A good number of thalassemic patients were on irregular treatment in regard to desferrioxamine or blood transfusion which make them more liable to get complications including bone changes as it is shown in table(2).

Table 2 Patients on desferrioxamine

Regularity	NO.	%
Regular	28	34.1
Irregular	52	65.9
Total	80	100

Measurement of wrist circumference of patients with splenomegaly compared with those without splenomegaly has shown a significant difference P<0.05and 59 healthy children were got their wrist circumference measured as a control for both groups of patients to avoid the possibility of age variation of wrist circumference between the two patients group and healthy children and the difference is highly significant P<0.001.

Table 3 The mean and standard deviation of wrist circumference in patients with and without splenomegaly and healthy children.

Group	NO.	Mean	SD	S.Error	95% Confidence interval for the mean Lower	Upper	P-Value
Patients with splenomegaly	52	11.9077	1.465	0.1451	11.6	12.2	< 0.05
Patients without splenomegaly	53	11.4358	1.047	0.1517	11.1	11.7	< 0.05
Normal children	59	10.3797	1.0015	0.137	10.110	10.650	< 0.001

Discussion

Our results show significantly the importance of wrist circumference as a measure of severity in comparison with splenomegaly and the confidence limit of the mean is highly predictive. Orofacial complications are a well known feature of widening of bones of thalassemia[18] and it indicates an advanced disease. Bones of the hand and wrist have been studied by CT scan and MRI[3] which has shown widening and sclerosis.X-RAY has been used in a study of the width of the metacarpals of thalassemic patients and it has been found wider than control group[19].The changes in bone width have been found to be due to bone marrow expansion[20]. Bone changes are found more prevalent in thalassemic patients with less transfusions and shorter desferroxamine courses which is true to our patients .Only 34.1% of our patients of both groups who have been put on desferrioxamine are on regular treatment due to poor compliance of these patients or unavailability of the drug which goes with the findings in thalassemia centre in Mosul and Saudi Arabia[21,22].

To my knowledge no study has been done to check wrist bones clinically by simply measuring the circumference in relation with splenomegaly to compare our results with.

Conclusion

Wrist circumference measurement in thalassemic patients is a simple, practical method to check bone changes and severity of the disease in comparison with splenomegaly especially in areas where sophisticated tests like MRI are not available.

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