ORIGINAL ARTICLE

The effect of sex steroids on pubertal disorders of beta thalassemia major.

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Abstract

Purpose: Pubertal disorders are well known Complications of beta thalassemia Major (TM.) . The aim of study was to assess the effect of sex steroid administration on thalassemic patient attending Boo Ali Sina Hospital, Sari, Iran.

Materials and Methods: all of the TM patients at Growth Clinic from 2005 to 2011 were underwent treatment with ethinylestradiol 2.5 to 25 μ g/ day plus medroxy progesterone acetate 5mg/day for 15 days of each month for females, or testosterone enenthate 50 –l00 μ g / month as IM injection for males. Weight, height, pubertal stage of the subjects was recorded before and after the intervention. Thyroid function, serum ferritin, Ca , P, ALP , LFT, OGTT tests were performed. Compliance to treatment measured by looking at missed visits.

Results: 173 TM Patients (54% Female) were followed, age at study in girls and boys were 14± 3.5 and 14.3 ± 2.5 years respectively. Follow up period for girls and boys were 3± 1.2 and 3.3±1.5 years respectively. Increasing of BW SDS and BH SDS in both sexes were statistically significant (P<0.01). SMR changes by McNemar test was significant only in girl (p<0.01). Compliance to treatment was better in girls.

Discussion: Sex steroids are cheap and available treatment for growth and pubertal disorders in TM patients. Compliance to treatment is essential for successful treatment.

Key words: Beta thalassmia major, puberty , Sex steroids, Treatment

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Introduction

Beta thalassemia major (TM) patients often have different types of pubertal disorders including: delayed or arrested puberty , primary amenorrhea, hypogonadism and even secondary amenorrhea or regression of pubertal achievements (1).

Short stature and low BMI are also associated and partially related to pubertal disorders (1,2) .It has been shown that all above mentioned disorders are common and sever a in our patients (3,4,5,6) .Different etiologies are reported to be important in the process including: malnutrition, chronic anemia, hyperslpenism , zinc deficiency, growth hormone deficiency, hypothalamus - hypophiseal -gonadal axis disorders(I)

Attention to and application of some of these factors are either difficult (because of problems in interpretation of the test results, expense and availability of treatment modalities) or non practical (because of low compliance of the patients). The objective of the present study was to investigate the effect of sex steroids on the growth and sex maturity of TM patients.

Materials and methods

Boo Ali Sina general hospital is an university center with a dedicated thalassmia ward and clinic. Till October 2011 , about 1000 thalassemics were registered in this ward and subspecialty clinics of the hospital. Since October 2005 all 12 years old and elder TM patients were regularly visited at the Growth Clinic. Growth of younger patients was checked in regular visits for blood transfusion. In Oct .2011 the growth clinic records were reviewed and the results of treatment of TM patients who were compliant to the treatment protocol for at least 1 year were reported. Body weight was measured using Seca scale and compared to the data of normal sex and age-matched population (Tanner- Whitehouse 1975 growth charts) and BW SDS was calculated. Height was measured using Serono stediometer and BH SDS was calculated in the same way. Sex maturity rating as Tanner stages were documented by experienced pediatric endocrinologist.

Follow up visits were performed every 3-4 months. Numbers and intervals of F/U visit were recorded to determine compliance to the treatment.

Thyroid function, serum ferritin ,Ca ,P, ALP, LFT and OGTT were measured by standard methods.

The study was fully explained to patients and they were given inform consent.

All the study protocols was approved by medical ethical committee of Mazandaran Medical University.

Paired T- test for quantitative variables and McNemar test for SMR changes were used.

Results

The records of 173 (45% female)TM patents were reviewed and reported. The age of the girls at the first blood transfusion and at the study time was 3.6±4.2(range : 0.1-30) years and 14±3.5 (range 12-30) years respectively and F/U period was 3±1.2 years. The age of boys at the first blood transfusion and at the study time was 3.5±2.8 (range 0.1-12)and 14.3±2. 5 (range 12-20)years respectively and F/U period was 3.3±1.5 years. Table 1 presents body weight and its SDS and body height and its SDS as well as SMR and also changes due to treatment. Increase of weight and height in the both sexes and SMR in girls were statistically significant ($P<0.01$) Compliance to the treatment was better in girls than boys who had frequent missed visits and interruption of treatments.

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	Female No.93		Male No:80	
	Before	**After**	**Before**	**After**
Weight(kg)	33.7±7.4	42.8±7.6	33.3±5.3	43.9±7.2
Height(cm).	139.8±10.6	148.6±7.7	140.9±9.3	154.8±9.7
BW SDS	-2.4±1.4	-1.9±1.4	-2.4±1.3	-2.4±0.93
BH SDS	-2.4±1.5	-1.9±1.2	-208±1.6	-2.6±1.3
T1	63(67.8)	1(1.07)	64(80)	14(17.5)
T2	15(16)	5(5.4)	14(17.5)	20(25)
T3	9(9.7)	21(22.6)	2(2.2)	17(21.3)
T4	2(2.2)	16(17)	0	17(21.3)
T5	4(4.5)	50(54)	0	12(15)
Ferritin (ng/m1)	1500±900	-	2400±1700	-

TABLE 1.

Discussion

At time of beginning of the study pubertal disorders were common and sever. Bazrafshan et al after an analytical study on 110 TM patients in Gorgan(Iran) showed that pubertal delay was command related to iron chelation status of their patients(5).Fatahi et al also reported growth and gonadal disorders in TM patients of Zahedan (Iran)(6).None of these studies presented the effect of treatment We previously have studied our patients during 1994-98 and found out that height improvement was significant but weight changes were not(8) .Those patients were elder, with lower BMI and pubertal delay was worse than the patients of this study. The duration of the treatment was also shorter in the previous review.

Other causes of short stature and pubertal delay such as hypothyroidism was not common in our patients and also overt hypothyroidism was rare (3, 9).

This study showed that without expensive and troublesome investigations and treatments we can improve the growth and pubertal situations of TM patients. Both of these two entities are very important regarding psychosocial and bone density of patients as well as general health of them (1, 10, and 11).

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