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Evaluation of Pulmonary Function In Young And Adolescent Patients With Beta- Thalassaemia.

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OBJECTIVES: We aimed at assessing lung function test in pediatric patients(8-18 yrs) suffering from thalassemia who are on blood transfusion and chelation therapy. METHODS: PFT was done to all the the subjects diagnosed with thalassemia and chest x-ray and echocardiography were done to excude overt cardiac failure.
RESULTS: FEV1, FVC, FEF25-75 were less compared to age and height-related predicted value. Of which reduction in FEF 25-75 was statistically significant. FEV1/FVC was increased and the result was also statistically significant. So, the results showed towards restrictive lung pathology.
CONCLUSIONS: Restrictive lung disease is a complication during the course of thalassemia which probably occurs due to iron deposition during the course of the disease itself or due to its treatment.

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I. Introduction

Beta thalassemia is one of the most common autosomal recessive disorders worldwide. High prevalence is present in population in the Meditteranean, Middle-East, Transcaucasus, Central Asia, Indian Subcontinent, and Far East . The highest incidences are reported in Cyprus, Sardinia, and South East Asia . Beta thalassemia is the commonest single-gene disorder found in Indian population. Ten percent of the total world thalassemics are born in India every day. More commonly affected communities are Sindhis, Gujratis, Punjabis, and Bengalis.

Patient with beta thalassemia major suffer from a qualitative defect in hemoglobin production, resulting in decreased oxygen delivery to the tissues, ineffective erythropoiesis, and iron overload. To improve the oxygen carrying capacity of the blood, these patients receive regular blood transfusion, resulting in more iron overloading. Iron deposition in the lungs has been observed on postmortem examination of patients who received multiple blood transfusion(1). Different studies on lung function test in beta thalassemia patients documented different types of lung injury(2). Some studies found pulmonary parenchymal disease, where others documented a restrictive defect or even an obstructive pattern(3), (4). This study was, therefore, conducted to determine the predominant lung dysfunction seen in beta thalassemia major patients.

II. Materials And Method

Patients evaluation:

We measured PFT in 42 patients with beta-thalassemia (21 M, 21 F, age range:8 to 18 years) enrolled from the Thalassemia Unit, CNMC, Kolkata from 3/12/2014 to 23/12/2014. To be enrolled into the study patients were required to be between 8 to 18 years of age, to be able to perform pulmonary function test and not to be in overt cardiac failure. The diagnosis of thalassemia was based on hematologic data (hemoglobin electrophoresis and complete blood count) provided by the college laboratory. To maintain a satisfactory Hb level all patients were receiving regular blood transfusion. 39 out of 42 patients were on oral chelation therapy with Deferoxamine. Among study subjects 18 had a splenectomy. No patients had a history of bronchial asthma or any allergic respiratory symptoms. All patients had clinical examination of upper and lower respiratory tract before undergoing PFT. Each one had a chest x-ray on entry to the study. FVC, FEV1, FEV1/FVC and FEF 25-75% were recorded using an electronic spirometer 24 hours before patients received planned blood transfusion. Cardiac functions were evaluated by ECG and echocardiogram: echocardiographic ejection fraction calculated from M- mode recordings or two dimensional M-mode studies were employed to assess cardiac function.

Statistical analysis:

Comparisons with normal values were made using Students unpaired t-test. Results were statistically significant when $p<0.05$. Pearson's correlation was used to analyze the joint effects of several variables. Summarized data are presented using correlation co-efficients and Mean +/- SD for group data.

III. Results

The study included 50 patients between 8 to 18 years who had been given a diagnosis of beta thalassemia , were randomly selected from the institution, whose physical characteristics are reported in Table 1. Of the selected patients 7 were excuded from the study for being developed signs of overt cardiac failure and 1for having signs of bronchopneumonia on chest x-ray. Table 2 showed main results of pulmonary functions in thalassemic patients. Results showed the trend towards restrictive lung changes in thalassemic patients. Results are expressed in Mean+/- SD. FEV1, FVC, FEF25-75 were less compared to age and height-related predicted value. Of which reduction in FEF 25-75 was statistically significant. FEV1/FVC was increased and the result was also statistically significant. Table 3 showed correlation between % changes in PFT parameters and different variables like serum ferritin, hemoglobin, age, number of transfusions and BMI. But we find no significant correlation between these variables.

Table 1: Baseline Characteristics of Study Population

Variable	N	Mean	Std. Dev.	Minimum	Maximum
Age (YRS)	42	12.12	2.77	8	18
Height (CMS)	42	124.90	10.52	101	151
Weight (KGS)	42	24.79	6.13	17	43
Chest Circumference(CMS)	42	65.35	4.34	60.5	77.5
Transfusion In Years	42	8.25	4.25	1	16.5
No of Transfusions	42	123.14	83.44	7	310

Gender Distribution

SEX		
SEX	**Number**	**Percent**
F	21	50
M	21	50

Distribution of Chelation Therapy

CHELATION THERAPY		
CHELATIONTHERAPY	**Number**	**Percent**
No	3	7.14
Yes	39	92.86

Distribution of Splenectomy

SPLENECTOMY		
SPLENECTOMYDONE	**Number**	**Percent**
No	24	57.14
Yes	18	42.86

Table 2: Comparison of Actual and Predicted PFT Parameters

Variable	Actual value	Predicted value	p
FEV1	1.16 ± 0.34	1.26 ± 0.36	0.196
FVC	1.30 ± 0.43	1.53 0.40	0.014
FEF 25-75	1.54 ± 0.49	2.1 ± 0.72	<0.0001
FEV1/FVC	91.22 ± 8.74	79.81 ± 3.64	<0.0001
PEFR	2.09 ± 0.71	2.28 ± 1.55	0.47

Values expressed as Mean ± SD. Significance assessed by unpaired t-test. $p<0.05$ is considered as statistically significant.

Table 3: Correlation between % change in PFT parameters and Different Variables

Pearson Correlation Coefficients						
Variable		**FEV % change**	**FVC % change**	**FEF 25-75 % change**	**FEV1/FVC %change**	**PEFR % change**
Serum ferritin	**r**	-0.19327	-0.17475	-0.15978	0.06189	-0.0739
	p	0.22	0.27	0.31	0.70	0.64
Hemoglobin	**r**	0.19314	0.17494	0.14902	-0.16365	-0.13755
	p	0.22	0.26	0.34	0.300	0.385
Age	**r**	0.11129	0.14103	-0.09287	-0.178	-0.2878
	p	0.48	0.37	0.56	0.25	0.064
No of Transfusions	**r**	-0.079	-0.03716	-0.2401	-0.04969	-0.18629
	p	0.62	0.82	0.12	0.75	0.23
BMI	**r**	0.17675	0.22314	0.08715	-0.14592	-0.0425
	p	0.26	0.15	0.58	0.35	0.78

$P < 0.05$ considered as statistically significant, r = correlation coefficient.

No significant correlation found.

IV. Discussion

This cross-sectional study was designed to see the effect of thalassemia on the lung due to iron deposition during the course of the disease. It was observed that the children suffering from the disease were also affected by restrictive lung disease. Should that problem diagnosed early we may reduce the complications and morbidity due to lung disease and thalassemia as well.

References

[1]. Cooper DM, Mansell AL, Weiner MA, Beadon WE, Chetty-Baktaviriam A, Reid L, et al. Lung capacity and hypoxemia in children with thalassemia major. Am Rev Respir Dis 1980; 121: 639-46

[2]. Witzleben CL, Wyatt JP. The effect of long survival on the pathology of thalassemia major. J Pathol Bacteriol 1961; 82: 1-12

[3]. Factor JM, Pottipati S, Rappaport I, Rosner IK, Lesser ML, Giardina PJ. Pulmonary function abnormalities in thalassemia major and the role of iron overload. Am J Respir Crit Care Med 1994; 149: 1570-74

[4]. Priftis KN, Anthrocopoulos MB, Tsakanika C,Tapaki G, Ladis V, Bush A, Nicolaidou P. Quatification of siderophages in bronchoalveolar fluid in transfusional and primary pulmonary hemosiderosis.Pediatr Pulmonol 2006; 41: 972-7

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