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ORIGINAL RESEARCH

A study on pulmonary function test among Children aged between 5-15 years with beta thalassemia major admitted for periodic blood transfusion in paediatric ward

[1]Dr.Bollakonda Maneesh Karthik, [2]Dr.Guruprasad B, [3]Dr. Mahendra Kumar Banakar

[1]Junior resident, Department of Pediatrics,KVG Medical College, Sullia, Karnataka, India
[2]Assistant Professor in Paediatrics, Department of Paediatrics, KVG Medical College, Sullia, Karnataka, India
[3]Assistant Professor in Paediatrics, Department of Paediatrics, KVG Medical College, Sullia, Karnataka, India

Corresponding Author
Dr. Mahendra Kumar Banakar
Assistant Professor in Paediatrics, Department of Paediatrics, KVG Medical College, Sullia, Karnataka, India

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ABSTRACT
Over the past three decades, regular blood transfusions and iron chelation have dramatically improved the quality of life and thalassemia is transformed from a rapidly fatal disease to a chronic disease and is compatible with prolonged life. Regular blood transfusion causes generalized iron overloading in organs such as heart, liver and pancreas. Lung impairment in thalassemia is also noted. All enrolled children were taken detailed clinical history including age at first blood transfusion, number of blood transfusion, duration of iron chelation therapy and General physical examination findings were recorded on a predesigned proforma. Total number of children with abnormal pulmonary function tests was 35(77.8%). Ten (22.2%) children had normal PFT. Among 35 children with pulmonary dysfunction, 31(88.5%) children had restrictive pattern, 2 (5.71%) children had Obstructive pattern and 2(5.71%) children had combined pattern. Mean age of children with abnormal PFT was 8.2± 2.4.

Key words: Pulmonary function test, children, thalassemia

INTRODUCTION

Thalassemia is derived from the Greek words "Thalassa" meaning seal and "haema" meaning of blood. Indeed, it was first described in people who lived around the Mediterranean by Cooley and Lee in 1925, and the name thalassemia was first used by Whipple and Bradford in 1932. Thalassemia is an autosomal recessive disorder and is the most common monogenic disorder worldwide. The condition is highly prevalent in Mediterranean countries, the Middle East, and Southeast Asia. However, due to widespread migration, thalassemia can now be found across theglobe[1].

Ten percent of the total world thalassemics are born in India every year[2]. In India prevalence of beta thalassemia is 1 to17% and carrier frequency is 3-4%[2].

Over the past three decades, regular blood transfusions and iron chelation have dramatically improved the quality of life and thalassemia is transformed from a rapidly fatal disease to a chronic disease and is compatible with prolonged life. Regular blood transfusion causes generalized iron overloading in organs such as heart, liver and pancreas. Lung impairment in thalassemia is also noted. However, lung dysfunction has never been adequately focused upon and remains to be one of the least understood complications. Although it does not produce any symptoms and is not the most significant clinical manifestation of thalassemia.

Most studies found that restrictive dysfunction is the predominant pattern of lung function abnormality, although some others found obstructive lung dysfunction and combined pattern also[3,4].The precise

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causes and pattern of pulmonary dysfunction in thalassemia has not yet been established.

Most of the studies have tried to correlate serum ferritin levels with pulmonary function abnormalities but the results are conflicting[5,6].

There are not many studies from our country especially from southern part of India, Hence we propose to take up this study of pulmonary function test in children with thalassemia.

METHODOLOGY

STUDY TYPE: Hospital based cross sectional study.

STUDY POPULATION: Children aged between 5-15 years with beta thalassemia major admitted for periodic blood transfusion in paediatric ward.

INCLUSION CRITERIA

Children with confirmed diagnosis of beta thalassemia major in the age group 5 years to 15 years.

EXCLUSION CRITERIA

1. Thalassemia children who were already diagnosed cases of pulmonary dysfunctions. (i.e. asthma, bronchiectasis and other chronic lung diseases).
2. Children with congenital heart disease/Rheumatic heart disease.

- Ethical Clearance was obtained from the Institutional Ethics Committee.
- Children who fulfill the inclusion/exclusion criteria for the study were selected.
- Informed and written consent was obtained from parents of all cases.
- All enrolled children were taken detailed clinical history including age at first blood transfusion, number of blood transfusion, duration of iron chelation therapy and General physical examination findings were recorded on a predesigned proforma.
- Before blood transfusion, for all enrolled children one blood sample was sent for Serum ferritin level and another sample for pre-transfusion Hb.
- Serum ferritin level was measured by electrochemi-luminescence technique using Cobas 6000 analyser.
- Pulmonary function test was done using spirometer (RMS Helios) (annexure1), within 24hrs of blood transfusion.

RESULTS

Table 1: Results of Pulmonary Lung Function Tests (N=45)

Results	Number (N=45)	Percentage (%)
Normal	10	22.2
Restrictive	31	68.9
Obstructive	2	4.4
Combined	2	4.4

Total number of children with abnormal pulmonary function tests was 35(77.8%). Ten (22.2%) children had normal PFT. Among 35 children with pulmonary dysfunction, 31(88.5%) children had restrictive pattern, 2 (5.71%) children had Obstructive pattern and 2(5.71%) children had combined pattern. Mean age of children with abnormal PFT was 8.2± 2.4. Among abnormal PFT (35 children), 20 children were male and 15 were female with M:F 1.3:1.

Table 2: Various Parameters of Pulmonary Function Tests (N=45)

PFT Parameters	Mean ±SD	Number of Cases (N=45)	
		Normal	Decreased
FEV1%	81.7±40.6	20(44.4%)	25(55.5%)
FVC%	68.9±20.6	12(26.7%)	33(73.3%)
FEV1/FVC%	121.4±39.8	41(91.1%)	4(8.9%)
PEFR	99.3±49.9	22(48.9%)	23(51.1%)
PEF25%-75%	82.4±31.6	22(48.9%)	23(51.1%)

The mean FEV1% was 81.7 ± 40.6, mean FVC% was 68.9 ± 20.6, mean FEV1/FVC% was 121.4 ± 39.8, mean PEFR was 99.3 ± 49.9 and mean PEF 25%-75% was 82.4 ± 31.6.

Twenty five (55.5%) Children had reduced FEV1 and 33(73.3%) had reduced FVC. However, 41(91.1%) had normal FEV1/FVC%, where as PEFR and PEF25%-75% was reduced in 22(51.1%) children each.

Table 3: Correlation between Age with Pulmonary Function Tests(N=45)

		Age	FVC%	FEV1%	FEV1/FVC%	PEFR
Age	**Pearson Correlation**	1	-0.435**	-0.324*	-0.081	-0.507**
	P value		0.003	0.03*	0.595	<0.001*

* Significant; ** Highly significant

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There was significant negative correlation between Age and FVC%, FEV1% and PEFR. i.e., with increase in age there was decrease in FVC%, FEV1% and PEFR.

Table 4: Comparison of Serum Ferritin with Respect to Pulmonary Lung Function Tests (N=45)

PFT Results	Serum Ferritin (ng/ml)		
	Mean	SD	Median
Normal	2868.70	2985.58	1610.90
Restrictive	3267.93	1995.89	3016.80
Obstructive	2792.40	2096.01	2792.40
Combined	1633.15	32.03	1633.15
P Value#	0.758		

Using ANOVA TEST

Mean Serum Ferritin among those with normal PFT was 2868.70 ± 2985.58, those with Restrictive pattern was 3267.93 ± 1995.89, those with Obstructive pattern was 2792.40 ± 2096.01 and those with combined pattern was 1633.15 ± 32.03. There was no significant difference in mean serum Ferritin with respect to diagnosis.

DISCUSSION

The pulmonary dysfunction in thalassemia major children can be restrictive, obstructive,andcombined.Impairmentinrespiratoryfunctionamongthalassemia children has been reported in the range of 29-86%(7-9,48,79). In our study 35(77.8%) children had abnormal pulmonary function test. Of these 35 children majority of them had restrictive pattern of lung impairment i.e., 88.5% (31 children) followed by 5.7% (4 children) obstructive pattern and 5.7% (4 children) combined pattern[7].

Restrictive lung function abnormality was observed as major pulmonary abnormality in our study. Several other studies have shown similar restrictive pattern as major pulmonary dysfunction. But none of them found exact aetiopathogenesis.

In the present study, age and height was inversely correlated with FVC%, FEV1% and PEFR. Our results was consistent with the previous literature, as reported by Abu-Ekteish *et al*.[4] They reported that the severity of restrictive abnormalities of thalassemic children was found to increase with age, which was also consistent with other previous publish reports.

In many other studies different aetiopathogenetic mechanism for development of restrictive lung dysfunction were reported such as, multiple blood transfusion, hypoxia, iron-overload, drug like desferrioximine., genetic structure.

However Gulhan *et al*.[8] found more number of obstructive dysfunction (46.2%). The probable mechanism underlying obstruction may be presence of bronchial dilatation and areas of air trapping that might indicate early changes of obstructive lung dysfunction.

We also observed the combined pattern in 4 children (5.7%). Similar finding was also observed in Said *et al*. 1(3.1%). The probable reason for small number of cases could be due limitations in screening.

In present study, the mean values of various parameters of pulmonary function tests were reduced. A reduced FVC% 33(73.3%), a reduced FEV1 25(55.5%), a normal FEV1/FVC ratio 41(91.2%), a reduced PEF25 %-75% 23(51.1%) were observed. Probably in our study pulmonary function abnormality may be partially explained by insufficient anatomic and functional development of lung during early infancy,4 as we observed age and height negatively correlated with various parameters of pulmonary function test. The other reason could be iron overload as serum ferritin (>2500ng/ml) had significantly associated with pulmonary dysfunction.

In our study we observed 23 (51.1%) children had reduced PEF25%-75%. Isolated reduction in PEF25%-75% indicates possibility of small air way obstruction, but in our study other parameters of PFTs were also reduced, hence, small airway obstruction was not considered in our study.

Although the measurement of serum ferritin is not the best quantitative estimate of body iron stores, thalassemic patients with a serum ferritin concentration of ≥3000ng/ dL have been reported to have a high probability of lung injury. Levels >2500ng/dL have been reported to be associated with a 4-fold higher risk of death.3 In present study,no correlation was found between serum ferritin levels and PFT. However, in our study FVC and FEV1, PEFR, PEF25%-75% values were found to be decreased in patients with a high ferritin level (>2500ng/dL) as compared with children with a low ferritin level (<2500ng/dL),butthesedifferenceswerestatisticallynotsignificant.Acomplexmechanism in addition to iron overload has been proposed to play important role in the development of lung dysfunction. But in some other studies have shown significant correlation between serum ferritin and PFT results. However, it is known that serum ferritin level changes during process of chelation, and do not necessarily reflecttotal body iron stores. There was no significant difference in mean values of age, BMI and pre-transfusion Hb with respect to Serum Ferritin levels.

CONCLUSION

- The pulmonary function test abnormality was observed in 35 (77.8%) children of beta thalassemia major.
- The most common pulmonary functional abnormality in order of frequency were restrictive

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31(88.5%), obstructive 4(5.7%) and combined 4(5.7%) pattern.

- We observed, a reduced FVC% 33(73.3%), a reduced FEV1 25(55.5%), a normal FEV1/FVC ratio 41(91.2%) and a reduced PEF25 %-75% 23(51.1%).

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