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ORIGINAL ARTICLE

Frequency of Fetal Hemoglobin Level in Siblings of Betathalassemia Major patients

NASIR RASHID[1], ALMAS RAZA[2], SYED ZEESHAN HAIDER NAQVI[3], SHEEMA KHAN[4]
[1,2,4]Assistant professor, Pathology department, Pak Red Crescent Medical College, Lahore
[3]Associate professor microbiology, Institute of microbiology and biotechnology, University of Lahore, Defence road campus, Lahore
Correspondence to Dr. Nasir Rashid, Assistant professor, Cell: 0321-4285189

ABSTRACT

Aim: To see the frequency of fetal haemoglobin level in siblings of beta thalassemia major (BTM) patients**.**
Study population: A total of 400 subjects were included.
Grouping: Group A includes normal siblings of BTM cases, B includes beta thalassemia trait (BTT) siblings of BTM cases and group C were healthy controls.
Selection criteria: Asymptomatic siblings of diagnosed cases of beta thalassemia major were included. Patients with history or diagnosis of any acute or chronic illnesses were excluded from the study. 5 ml of blood was taken in EDTA vial and used for haemoglobin electrophoresis.
Results: In 200 cases, 78% (156) were beta thalassemia trait (BTT) siblings and fetal Hb was also more in BTT siblings i.e. Group B.
Conclusion: Fetal haemoglobin (HbF) was increased in BTT siblings (group B) of BTM cases.
Keywords: Hb electrophoresis, fetal Hb, beta thalassemia major

INTRODUCTION

Beta thalassemia usually produces marked anaemia in their homozygous and compound heterozygous states[1]. Their types are thalassaemia major, intermedia and minor[2]. The homozygous form i.e. thalassemia major is a serious problem with severe anemia, hemolysis, size/shape of the RBC variation, increased HbF, erythroblastosis, jaundice and hepatosplenomegaly[3]. The children develop thalassaemic facies and there is thinning of cortex of bones and due to this there is a tendency to fractures[4].

The objective of the study was to see the frequency of fetal haemoglobin level in siblings of beta thalassemia major (BTM) patients**.**

METHODOLOGY

Study Place: Thalassemia centres of SGRH Lahore, IBTS, Children Hospital and Services Hospital, Lahore.
Sample size: 400 subjects
Groupings: Three groups i.e. group A having normal siblings of BTM patients, group B having BTT siblings and Group C were healthy controls.
Study design: Cross sectional descriptive study
Data Collection: 200 cases of BTM were included and were brothers and sisters with two hundred healthy control group. Consent was taken and data e.g. age, gender, brief clinical history was taken. Five ml of blood in EDTA vial was taken and Hb electrophoresis was done. Approval was obtained from Institutional Ethical Review Board to start this study.

RESULTS

Table I: Distribution of ages in groups

Groupings	Mean ±Sd	Ranges	Total
A (Normal siblings)	11.3±6.6	1-26	44
B (BTT siblings	11.8±6.9	1-30	156
C (Controls)	10.6±6.5	1-29	200

AvsB=p<0.05 (significant) AvsC=p>0.05 (Non significant)
BvsC=p<0.05 (significant)

Table II: Gender distribution

Gender	A	B	C
Male	27(61.3%)	89(57.1%)	108 (54%)
Female	17(38.7%)	67(42.9%)	92(46%)
Total	44 (100%)	156(100%)	200(100%)

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Table III: Comparison of Fetal Hb in groups A, B and C

Fetal Hb	Mean ± SD	Ranges	Total
Group A	0.72±0.12	0.3—0.8	44
Group B	2.1±0.7	0.4-3.4	156
Group C	0.91 ± 0.7	0.2-0.9	200

AvsB=p<0.05 (significant)
AvsC=p>0.05 (Non significant)
BvsC=p<0.05 (significant)

Table IV: Siblings of BTM patients

	n=	%age
Group A	44	22%
Group B	156	78%

DISCUSSION

In this study, 156 (78%) cases were diagnosed as beta thalassaemia trait (BTT) and 44(22%) cases were normal siblings. In another study in Bombay, frequency was 48% among the siblings of BTM patients in total of 200 cases[5]. This study is in favour of our study. Thalassaemia trait in Pakistan is about 5.4%. BTM is common in pathan population i.e. 7.96% than Punjabi population (3.3%). The high incidence in pathans may be due to consanguineous marriages[6].

In one study, ages in beta thalassaemia heterozygotes ranged from 6 months to 86 years[7]. M:F ratio was 1.4:1 in the siblings of thalssaemia major and in the controls was 1.2:1. M:F ratio in the beta thalasaemia trait detected during the present study was 1.3:1. It has been observed that males are more prone to develop BTT as compared to females.
Mean± SD value of HbF in present study was 2.1±0.69% (0.6—6.6). These results are in agreement with (zero to 7.8%) by Pootrakul et al[8] and (1--6.8%) by Alter[9].

CONCLUSION

Fetal haemoglobin (HbF) was increased in BTT siblings (i.e. Group B) of BTM cases.
Conflict of interest: Nil

REFERENCES

1. Forget BG, Olivieri NF. Hb synthesis and the thalassemias. In: Handin R1, Lux SE, Stossel TP, eds. Blood principles and practice. 2nd ed. Philadelphia: Lippincott Williams e Wilkins, 2003: 503-96.

2. Ahmed S, Saleem M, Model B et al. Screening; extended families for genetic Hb disorders in Pakistan. N Engl J Med 2002; 347: 1162-8.
3. Brittenham GM, Cohen AR, McLaren CE. Hepatic iron stores and plasma ferritin levels in patients with sickle cell anemia and thalassaemia major. Am J Haematol 1993; 42:81-85.
4. Hoffbrand AV, Pettit J, Moss PAH. Essential Haematology 5th Ed. Oxford: lack well Scientific Publiscation 2006: 72-93.
5. Saenger P, Sehwartz E, Markenson AL et al. Depressed serum somatomedin activity in β-thalassaemia. J Paed 1981; 9(2): 214-18.
6. Khattak ME, Saleem M. Prevalence of Heterozygous beta-thalassemia in Northern areas of Pakistan. J Pak Med Assoc 1992; 42: 32-4.
7. Clegg JB, Weatherall DJ. Molecular basis of thalassaemia. Br Med Bull 1976; 32(2): 262-69.
8. Pootrakul P, Wasi P, Nakorn SNA. Hematological data in 312 cases of BTT in Thailand. Br J Haematol 1973; 24: 703-12.
9. Alter BP. Prenatal diagnosis of hemoglobinopathies and other hematological disorders. J Paed 1979; 95 (4): 501-13.