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Original Research Article

Study of distribution of ABO blood groups in ß-thalassemia patients

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ABSTRACT

Background: ß-thalassemia is an inherited hematological disorder caused by reduced or absent synthesis of ß globin chains of the hemoglobin which causes hemolysis and impair erythropoiesis. Affected children thus require regular lifelong blood transfusions. Blood group is one of the important and comparatively known parameter today which exhibits a strong correlation with some common diseases like cardiovascular diseases, gastric cancer, HIV infection etc. Aim of this study hence was to study relation between ABO blood groups and ß-thalassemia and to study complications in each blood groups to make it easy to predict the type of population which is more prone or resistant to ß-thalassemia for understanding the regional demand of rare blood groups and to tide over the demands.
Methods: This was a cross sectional, single centric, open labeled, observational study in which 100 patients of ß-thalassemia were screened for their ABO blood groups after permission from institutional ethics committee and informed consent from the patient.
Results: Most common blood group in ß-thalassemia patient is O +ve. Post-disease complication is commonly seen with B +ve. Early onset of beta-thalassemia is seen in A +ve and B +ve. Late onset is seen with O +ve. Severity more with B –ve while O +ve shows disease in less severe form.
Conclusions: On analysis, it was found that the most common blood group getting affected by the disease ß-thalassemia is O +ve with the same people having higher chances of family history of the same disease. Within the family members who have the positive history of the disease, most common was O +ve blood group again.

Keywords: ß-thalassemia, Blood groups, Post-disease complication

INTRODUCTION

The thalassemias (Greek: thalassa-meaning sea) are a group of single gene inherited autosomal recessive hematological disorders caused by defects in the synthesis of one or more of the hemoglobin chains that cause hemolytic anemia.[1,2] α and ß-thalassemias are caused due to reduced or absent synthesis of α and ß globin chains respectively. The imbalance of the globin chains causes hemolysis and it impairs erythropoiesis.

Phenotypically ß-thalassemia is of three types. ß-thalassemia minor is a heterozygous state in which there is around 50% decrease in synthesis of ß-globin protein, causing mild to moderate microcytic anemia. Affected individuals are usually asymptomatic. ß-thalassemia intermedia shows mild to moderate anemia.

Most cases do not need blood transfusions. ß-thalassemia major is the most severe form which causes hemolytic anemia, poor growth, delayed sexual maturation and skeletal abnormalities during infancy.[1] Symptoms of the disease appear in first year of life as HbF synthesis reduces. Complications that occur due to ß-thalassemia are due to overstimulation of bone marrow, ineffective erythropoiesis. Pallor is usually the first sign

accompanied by splenomegaly of various severity, fever and failure to thrive.[2]

The diagnosis can be made by complete blood count (CBC), Hb-electrophoresis, DNA testing. Prenatal diagnostic tests include: chorionic villus sampling, amniocentesis, fetal blood sampling, fetal cell analysis in maternal blood.[3] Blood group is one of the important and comparatively known parameter to the large number of present population which exhibits a strong correlation with some common diseases like cardiovascular diseases, gastric cancer, HIV infection and more to enumerate.

This study is thereby an attempt to explore any relationship between blood group antigens and ß-thalassemia so that it will become very easy to predict the type of population which is more prone or resistant to ß-thalassemia. Also, the study of this relation between blood groups and the disease is extremely important for understanding the regional demand of rare blood groups and to tide over the massive demands.

The objective of this study was to study relation, affinity and family predisposition between ß-thalassemia and specific blood groups and to study the complications in terms of frequency, severity and onset in such patients in specific blood groups.

METHODS

This was a cross sectional, single centric, open labeled, observational study performed in department of physiology, Grant government medical college Mumbai, Maharashtra, India in collaboration with thalassemia unit of St. George hospital, Mumbai, Maharashtra, India during a period of August 2016 to October 2016. Which included total 200 participants: cases (100) registered patients of ß-thalassemia (67 males and 33 females) (between 5 years to 35 years) and controls (100) normal subjects (42 males and 58 females) (between 5 years to 35 years).

After approval from institutional ethical committee of Grant government medical college and Sir J.J. group of hospitals Mumbai, Maharashtra, India and informed consent from the patient. Data compiled in a questionnaire based on recorded history, family history and complication history. Blood group determined by Slide method in the department of physiology Grant government medical college, Mumbai, Maharashtra, India.[4] Results prepared in tabular and graphical format in Microsoft excel 2013.

Statistical analysis

Data about thalassemia related complications was analysed using Fisher's exact t test. p value 0.05 is considered significant and less than 0.01 is considered statistically highly significant.

RESULTS

It was found that among the study group of 100 cases, maximum were O +ve (39%) followed by B +ve which was about 29% of the cases studied. While in those who were selected as controls, 36% i.e. maximum were the people with B +ve blood group. The overall percentage distribution is represented in the Table 1:

Table 1: ABO-blood group distribution in the ß-thalassemia patients (cases and controls).

Blood Group	Cases (n=100)	Controls (n=100)
A +ve	17	17
A –ve	3	3
B +ve	29	36
B –ve	2	3
AB +ve	8	9
AB –ve	1	2
O +ve	39	29
O –ve	1	1

Statistically comparing the data about the thalassemia related complications within +ve and –ve blood groups of the same ABO blood group, no significant result was obtained. Which states that there is no specific predisposition of complications in relation to presence or absence of Rh factor. Fisher's exact t test was applied for the same.

The result of data concerned with onset of complications in individual blood group revealed that earlier onset of complications (i.e. within 5 years of life) was observed in A +ve and B +ve blood groups. Higher incidence of earlier complications was however seen with B +ve to A +ve. While late onset of complications was recorded with O +ve blood group being about after the age of 15-20 years of life. Tabular record of the related data is shown in Table 2.

Table 2: Onset of the thalassemia complications in individual blood groups.

Blood groups	Within 5 years	5-10 years	10-15 years	15-20 years	>20 years
A +ve	1	1	None	None	None
A –ve	None	None	None	None	None
B +ve	1	2	8	3	1
B –ve	None	None	None	None	None
AB +ve	None	None	None	1	None
AB –ve	None	None	None	None	None
O +ve	None	None	2	None	2
O –ve	None	None	None	None	None
Total	2	3	10	4	3

The data collected for recording the frequency of complications based on occurrence of the disease complications (fever, jaundice, pain in abdomen or the

transfusion complications like transfusion reactions, Hepatitis-C, Hepatitis-B infections) occurred how many times till the date from the beginning of regular blood transfusion, reported that the higher frequency of complication (i.e. 3 times) was seen with O +ve only. Also, the once and twice occurrence of complications accounted in O +ve cases to support the statement that O +ve shows highest frequency of complications. While no incidence of complications was seen with B –ve. Other frequencies have been mentioned in the table.

The days of hospitalization for the management of disease complications was taken as a criterion to estimate severity of the disease in individual blood group. We found that, B –ve was the blood group which needed maximum days (more than 7 days) of hospitalization in maximum number of patients to manage the complications. While O +ve was the blood group which could manage the complication even without hospitalization indicating less severe form of the complications in the blood group.

DISCUSSION

From the Table 1, it can be concluded that the most common blood group observed in patients of ß-thalassemia is O +ve in 39% of the patients. With 36% B+ve becomes the common blood group in the normal population. AB-ve and O –ve people are less likely to get affected by the disease. Comparing it with common population, O –ve becomes a blood group presenting in lesser number of people. The details collected for family history of the patients and the blood group of those affected relatives showed that more than 1/3rd of the patients have a positive family history. And from the data collected it was found that O+ve is a blood group common among the relatives too who were affected with the disease.

On comparing complication history among the +ve and –ve blood groups Table 3 concluded that there is no significant distribution amongst the two in relation to complication distribution. Table 2 concludes that of all the cases in the study, B +ve have maximum history of complications followed by O -ve. Least incidences of complications are seen with A +ve. Most common post-disease complication was anemia due to sequestration. Splenectomy was done in most of the population to avoid the complications or due to complications.

Table 3: Comparison of complications in the same +ve and –ve Blood Groups.

Blood group	Complications seen	Complications not seen	P-value
A +ve	3	14	0.5^{NS}
A –ve	1	2	
B +ve	12	17	0.51 ^{NS}
B –ve	0	2	
AB +ve	2	6	1.0 ^{NS}
AB –ve	0	1	
O +ve	16	23	0.4 ^{NS}
O –ve	1	0	
Rh +ve	33	50	0.7 ^{NS}
Rh –ve	2	5	

*P<0.05, **P<0.001, NS = not significant.

Table 4: Frequency of complications of thalassemia in individual blood groups.

Blood groups	Once	Twice	Thrice
A +ve	2	2	None
A –ve	None	1	None
B +ve	9	3	None
B –ve	None	None	None
AB +ve	1	1	None
AB –ve	None	None	None
O +ve	10	3	1
O –ve	None	1	None
Total	22	11	1

Table 5: Days of hospitalization for management of the disease complication (severity).

Blood groups	No hospitalization	1-2 days	3-6 days	7-10 days	>10 days
A +ve	1	None	None	2	1
A –ve	None	None	None	None	1
B +ve	3	None	5	None	None
B –ve	None	None	None	None	4
AB +ve	None	None	None	1	1
AB -ve	None	None	None	None	None
O +ve	7	1	None	3	4
O –ve	None	None	None	1	None

Followed by Hepatitis C being most common complication due to transfusions. Higher frequency of complications is seen with O -ve patients, B -ve is the blood group with the lesser frequency of complications,

according to Table 4. These complications were found to occur as early as by 5 years of life in the patients with A +ve and B +ve blood groups. People with O +ve blood groups are shown to experience the complications at the later stages of life as per the analysis of Table 5. O +ve patients conquering the complications needed no hospitalization at all to manage the complications in most of the cases indicating less severity of the complications in them. While B -ve were the people who needed long term, hospitalization showing higher severity of complications in the blood group.

Saha AK, et al found in their study of frequency and distribution of blood groups in the donors of a rural hospitals, West Bengal also got the finding of blood group B being common in blood donors.[5] Iqbal M et al, seen frequency of ABO and Rh blood groups in healthy donors had reported the same finding that B blood group is common among the donors.[6] While Mohammadali F et al, reported O +ve blood group to be common in the research of association of ABO and Rh blood groups to blood borne infections among blood donors in Tehran-Iran.[7] Mohssin MY et al, studied of frequency distribution of hemoglobin variant and ABO blood groups among thalassemia patients from Ibn-Al-Baladi hospital in Baghdad/Iraq stated the same fact of O blood group being common incidence (59.1%) and AB with the least common occurrence of the disease ß-thalassemia.[1] Ansari H et al, found prevalence of hepatitis-C and related factors among ß-thalassemia major patients in Southern Iran which stated that HCV rate in ß-thalassemia patients is seen more in patients of blood group O which is in favour of our finding which stated that complications of the disease are seen more common in O blood group of which HCV infection is the common complication shown in Figure 1.[8]

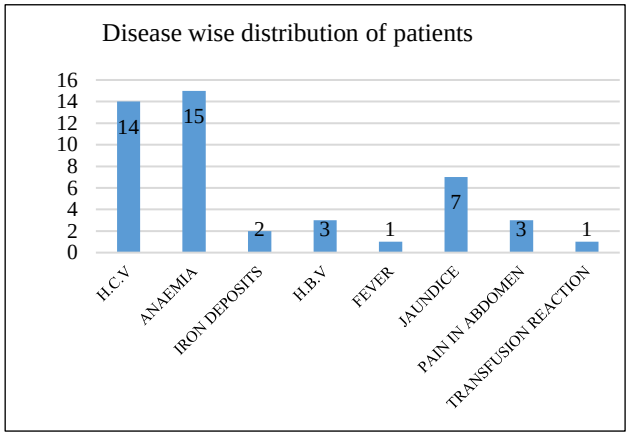

Figure 1: Different complications of the disease in the study cases.

Conventional treatment for ß-thalassemia includes regular lifelong blood transfusions from early childhood which improves anemia and reduces skeletal deformities associated with excessive erythropoiesis.[9] Although blood transfusion is life saver major, it is not the curative treatment. The only available curative treatment is bone marrow transplantation which is not feasible due to the complications and high cost.[10] Also, blood transfusion is not always safe, sometimes it may lead to dangerous reactions, the transfusion reactions.[11] Hence, pre-transfusion ABO and Rh blood group testing becomes mandatory.[12] Blood transfusion has many other associated complications too like iron overload, transfusion transmitted infections. Iron is deposited in visceral organs (heart, liver and endocrine glands).[13] The gastro-intestinal manifestations include nausea, vomiting, diarrhea, constipation etc.[4] Iron deposition hence demands iron chelation treatment from starting from around 5 to 8 years of age. Deferoxamine subcutaneously or intravenously is the treatment. Endocrinopathies (hypogonadism and diabetes mellitus) can occur in adolescents and adults due iron deposition in endocrine glands. Repetitions of transfusions provoke patient's immune system and produces anti-erythrocyte antibodies.[14,15] Splenomegaly may worsen the anemia present, also causes neutropenia and thrombocytopenia. Thromboembolic events may occur. Thalassemia traits have a normal life expectancy while ß-thalassemia major usually live a life of 30 years on an average. Major often die from cardiac complications of iron overload.

CONCLUSION

On analysis, it was found that the most common blood group getting affected by the disease ß-thalassemia is O +ve with the same people having higher chances of family history of the same disease. Within the family members who have the positive history of the disease, most common was O +ve blood group again. Complication history was common with O +ve people. HCV was the most common disease found as a complication of the disease management. Early onset of these complications was seen with A +ve and B +ve. Late onset in O +ve. Severity wise B –ve were found to be having the disease in more severe form while O +ve presenting the complications in lesser severe form. A study of thalassemia disease, its progress, complications and management options shows that managing a thalassemia patient is a lengthy process and is tedious for the patient. Hence the target of management of this disease should ideally be the prevention of the disease. Hence this study was planned to study the blood group distribution, frequency of the complications and study of complications in each so that it can be applied over the population to know who are to be counselled more about this disease and whom are we concerned more for pre-conceptional tests for the disease.

Hence, it is advisable to do the study over a large population and considering more parameters for applying the findings over the entire population. Detailed blood group wise study should be performed in which sample size will be equal in all blood groups. ß-thalassemia minor/intermedia and α-thalassemia should also be

studied for their correlation with the blood group and further outcomes of the disease.

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