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Current Situation of Thalassemia in Paktia Regional Hospital, Paktia, Afghanistan

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Abstract

Background*: Thalassemia is the most common hereditary disorder in the world including Afghanistan. The thalassemia is a major health problem all over the world, but this is particularly in the developing countries.*
Objectives: *The present study aimed to determine the Current situation of thalassemia in Paktia Regional Hospital during the 21 months of 2019 and 2020.*
Methodology: *This is a descriptive study conducted in Paktai Regional hospital in Paktia among patients*
diagnosed with thalassemia who require regular blood transfusions. Statistical analysis: The statistical analysis was performed by using the EPI INFO software package 6.04 version. The percentages were calculated for the various parameters which were under study.
Results: *Total of 137(100%) cases of thalassemia were included in the study. Amongst them 71 (51.82%) were males and 66(48.17%) were females. The majority cases 90(65.69%) were form urban area and the remaining 47(34.31%) were from rural area especially closed district. The majority of the cases were belonging to the younger 0-6 years of age group 105(76.64%)*
Conclusion: *Since thalassemia is most common monogenetic inherited disease, and leads to fatal complications. Therefore, the data obtained from this study can be used by the public health authorities, to prepare a suitable action plan for reducing the burden of the disease and providing treatment and support to people already suffering from it. Families having thalassemic child should be educated about the disease. All members of the family should be screened for thalassemia trait and genetic counselling should be done.*

Keywords: Thalassemia, Paktia, Current Situation**.**

Introduction

Thalassemia is the commonest monogenic disorder in the world that result from absence of or decreased globin chain
production. (1) The two main types of thalassemia are alpha and beta : Individuals with alpha thalassemia don't produce enough alpha globin chains causing excess beta globin chains; those with beta thalassemia don't produce
enough beta globin chains, causing excess alpha chains.The common signs and symptoms of thalassemic diseases include pale skin, retarded growth & puberty, anemia, enlarged spleen, and increased susceptibility to
infections.(2)High prevalence of Beta-Thalassemia is present in Mediterranean, Middle-East, Transcaucasia,
Central Asia, Indian subcontinent, and Far East(3). Thalassemia can be controlled by preventive health services.
Education about the risks of conceiving a child with thalassemia combined with family planning services is an effective approach to inform the behaviors of those at risk. Laboratory blood tests can diagnose the status of thalassemia in a person. Screening and genetic counselling are associated with numerous benefits, including
decreased number of thalassemic newborns. (4)

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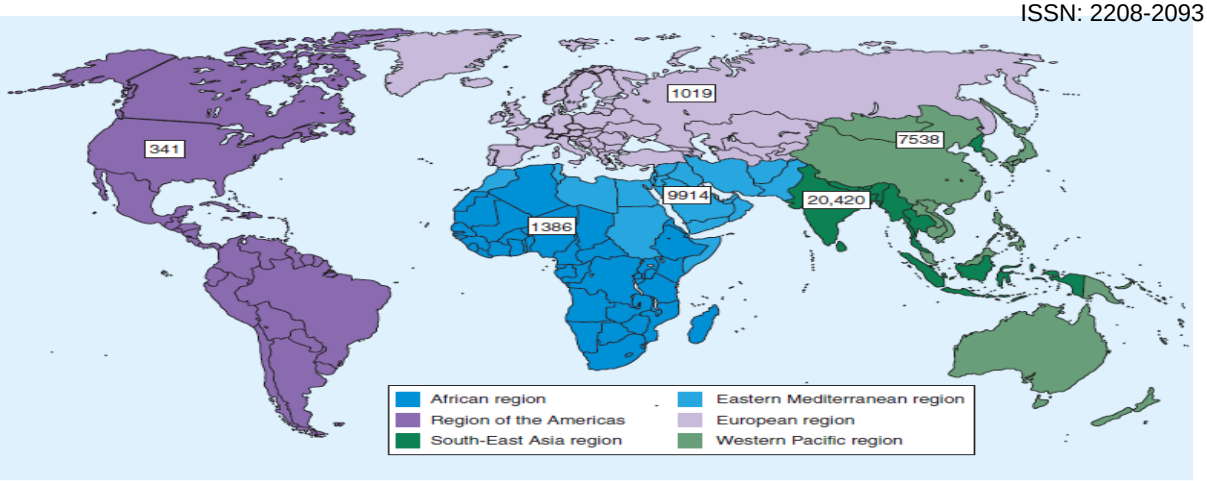

Figure (1-1) Colah, R., Gorakshakar, A., & Nadkarni, A. (2010).

Around 7% of the global population carries an abnormal hemoglobin gene, 300,000-500,000 children are born with clinically significant hemoglobin disorders annually , About 80% of affected children are born in developing countries , About 70% are born with Sickle Cell Disease and the rest with Thalassemia Syndromes , 50-80% of children with SCD die each year in low and middle income countries and 50,000-100,000 children with thalassemia major die each year in low and middle income countries(5) Transfusion therapy is the mainstay of management of thalassemia major. Since thalassemia is growing in populations globally, it is estimated that there will be around 900,000 new clinically significant thalassemia patients by the year 2025 (6) Transfusion therapy is life support for severely anemic thalassemic patients, but it comes with its side effects, the most prominent being iron overload related complications. Not only the amount of
blood transfused but also the duration of iron exposure gives the best estimate of organ dysfunction as observed in the study which showed iron overload cardiomyopathy, increased liver iron, and endocrine organopathy in transfusion-dependent thalassemia patients which were due to iron deposition in these organs.(7)
Parental awareness regarding various aspects of beta thalassaemia is of great importance not only for the proper management and improved quality of life of the patient with thalassemia but also for the prevention of further children with thalassemia major in the family (13).

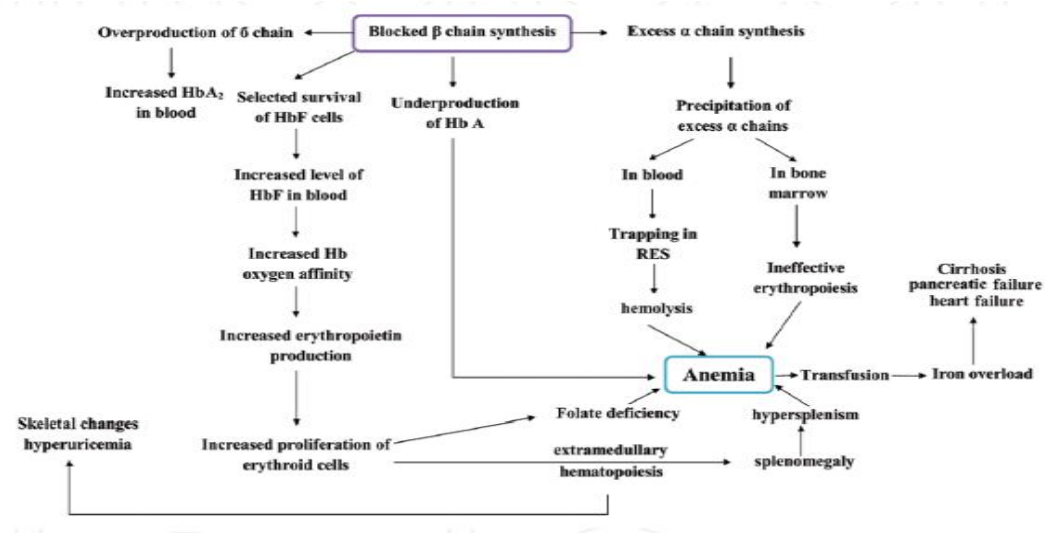

Figure 1.2: pathophysiology of beta thalassemia

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Thalassemia diseases are classified into transfusion-dependent thalassemia (TDT) and non-transfusion-dependent thalassemia (NTDT). This classification is based on the clinical severity of patients determining whether they do need regular blood transfusions to survive (TDT) or not (NTDT) (14)

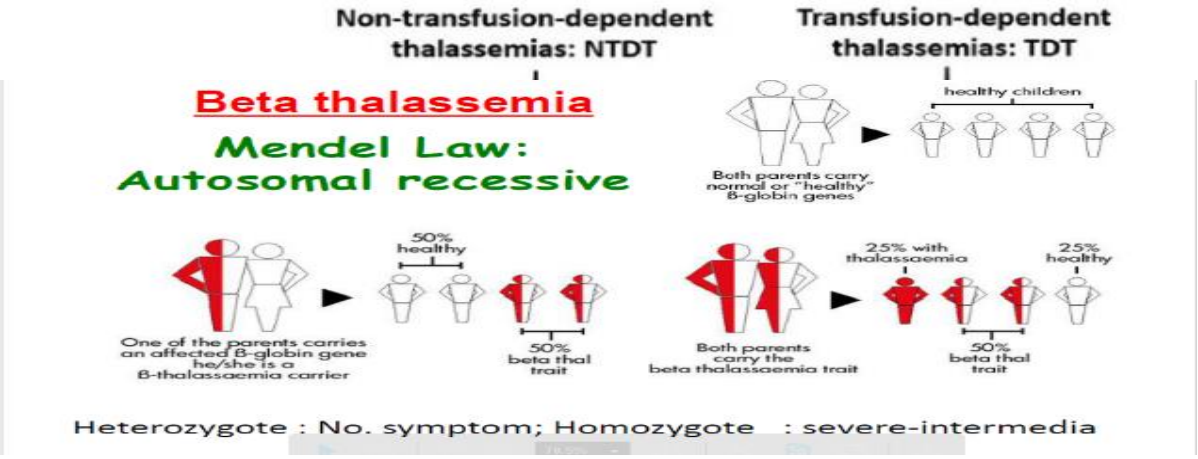

Figure (1-3): The clinical spectrum of thalassemia syndromes based on their requirement of regular blood transfusions into non–transfusion dependent thalassemia (NTDT) and transfusion dependent thalassemia (TDT).

Figure (1-4): Mendel law for beta thalassemia

Methods

Study type and study setting: This was an institution based observational descriptive study, descriptive in design, carried out in a tertiary regional hospital in Paktia, Afghanistan during the 21 months period from 2019 to June 2020. Study population was patients attended general OPD of Paktia Regional hospital on the months of 2019 to 2020. Inclusion criteria were aged 18 and below, both sexes, not seriously ill, gave informed oral consent to participate in the study. Exclusion criteria were aged above 18years, seriously ill, not gave informed oral consent.

Results

Total of 137(100%) cases of thalassemia were included in the study. Amongst them 71 (51.82%) were males and 66(48.17%) were females. The majority cases 90(65.69%) were form urban area and the remaining 47(34.31%) were from rural area especially closed district. The majority of the cases were belonging to the younger 0-6 years of age group 105(76.64%). According to the treatment, thalassemic children were taking only blood. And pre diagnosed, conformed thalassemic children were registered and were taking blood there in hospital. Awareness regarding different aspect of thalassemia were less among all subject, their parents and relatives.

[Tab/Fig-1]: Age and sex distribution of study subjects

Age (years) Groups	**gender**		**total**
	male	female	
0-6	55	50	105
6.1-10	12	14	26
Above 10	4	2	6
Total	71	66	137

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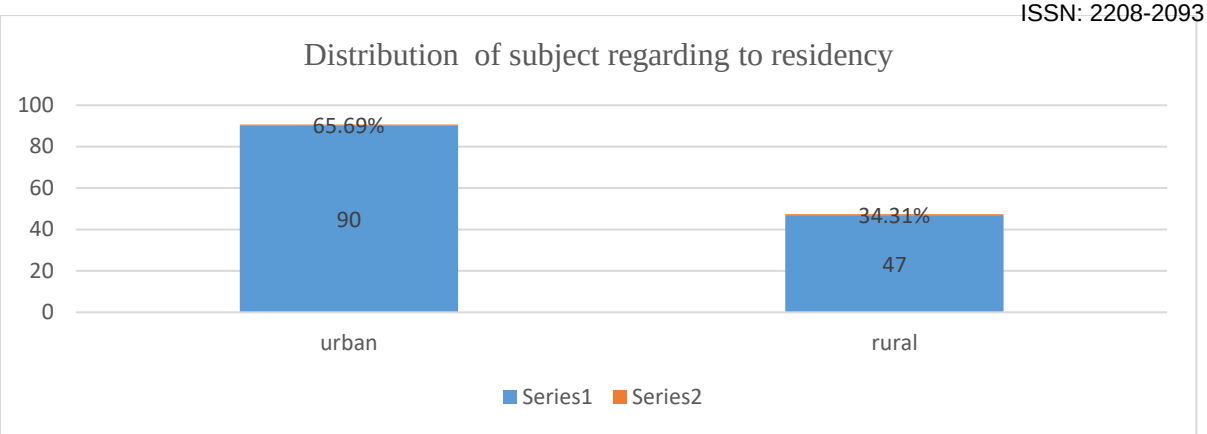

[Tab/Fig-2]: Distribution of all subject regarding to their residency

Discussion

In the current study among the 137 study participants, the numbers of thalassemic males (51.82%) were more as compared to females (48.17.3%) in the affected families. Other studies have found Similar, male preponderance was reported by studies from Zahidan southeast Iran male: female proportion (54.3:45.7) %(8) and India Ahmad Abad (67:33) %.(9) Current study showed that regarding to residency type rural area participant were 34.31% more affected than urban (65.69%). The result of this study is different to a study which was conducted in Shiekh Zayed medical college Karachi Pakistan there was difference in rural and urban area spread of disease 87(31%) belonged to the urban areas and 196 (69%) were resident of the rural areas (10). In our study majority of participants 105(76.64%) were in age group 0 to 6 years; which is supported by other studies like Sheikh Zayed medical college Lahore Pakistan (11) and India(12) the same result the majority of the cases were belonging to the younger 0-5 years of age group (55%). And 69.5% in age zero to 5 years respectively.

Regarding to our finding we recommend below points.

- ✓ There should be a national program for thalassemia control. MoPH should work on registration, prevention, prenatal detection, information and supportive programs. As a major public health issue.
- ✓ Improve registration and diagnosis of thalassemic children at Paktia regional Hospital. New patients need to have a comprehensive registration process and a proper electrophoresis diagnosis.
- ✓ Medical consultation program implantation is suggested, which includes patients' visits and monitoring complications for heart problems, growth, splenomegaly and infections.
- ✓ Looking for iron overload with serum ferritin, TIBC test and treating with iron chelating agent*+ (deferoxamine) should be provided in the pediatric ward thalassemia unit.

Conclusions

This study showed that males were suffering from thalassemia disorder in higher number as compared to the females. All these figures are consistent with national and international studies. In present study, the rural-to-urban prevalence ratio for beta-thalassemia was 3:1. This ritual should be discouraged especially in thalassemia families. Serum ferritin levels also need attention and iron chelation therapy should be encouraged. Families having thalassemic child should be educated about the disease. All members of the family should be screened for thalassemia trait and genetic counselling should be done. Moreover, in order to minimise blood transfusions in thalassemic patients and to cure the disease.

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