International Journal for Multidisciplinary Research (IJFMR)
E-ISSN: 2582-2160 ● Website: www.ijfmr.com ● Email: editor@ijfmr.com

Social Impact of Thalassemia: Assessing Social Well-Being and Quality of Life

Mr. Akhilesh Kumar Yadav[1], Ms. Kirti Kumari[2]

[1]Research Scholar, Pharmacy, Himalayan Institute Of Pharmacy And Research
[2]Assistant Professor, Pharmacy, Tulas University

Abstract
A genetic blood condition called thalassemia is linked to a decrease in hemoglobin in red blood cells (RBCs), which in turn results in less oxygen getting to the body's cells. Alpha-globin and beta-globin, components of hemoglobin, the protein that carries oxygen in the blood, are produced defectively in thalassemia, a hereditary condition. The Greek terms "Thalassa" (sea) and "Haema" (blood) are the origin of the name. Alpha-thalassemia and beta-thalassemia are the two primary forms. The Thalassemia National Federation observed Thalassemia Day for the first time on May 8, 1994.
The study found that children with moderate alpha-thalassemia are naturally protected from malaria-related anemia. Eight hundred children participated in the study, which was carried out in Papua New Guinea. In addition to the high prevalence of alpha-thalassemia in youngsters, malaria is endemic in this area. Globally, thalassemia is common; in 2013, it caused 25,000 fatalities. The Mediterranean, Italy, Greece, Turkey, West Asia, North Africa, South Asia, and Southeast Asia have the highest rates. Children with β-thalassemia major, the most severe type, must get frequent blood transfusions to survive.

Keywords: Thalassemia, Hemoglobinopathies, Bone Marrow Transplant, Gene Therapy, Gene therapy, Hypoxia

INTRODUCTION

Alpha-globin and beta-globin, components of hemoglobin, the protein that carries oxygen in the blood, are produced defectively in thalassemia, a hereditary condition. The Greek terms "Thalassa" (sea) and "Haema" (blood) are the origin of the name. Mutations in the genes that produce these globin chains, which are found on chromosomes 11 (beta) and 16 (alpha), cause this hereditary disorder. Alpha-thalassemia and beta-thalassemia are the two primary forms. In extreme situations, the illness causes severe anemia, which, if left untreated, can be lethal. The severity of the symptoms varies according on the number of globin chains that are impacted. Babies with severe beta-thalassemia, for example, typically don't exhibit symptoms until four to six months after birth. While lower variants of alpha-thalassemia allow for survival, the most severe form causes pregnancy difficulties. The severity of thalassemia may also be influenced by additional genetic factors, such as co-inheritance of other blood disorders or mutations that alter fetal hemoglobin production.
This genetic disorder is most prevalent in Mediterranean nations. From lesser and asymptomatic to severe and potentially fatal, the resulting anemia can manifest in a variety of ways. Splenomegaly and bone marrow growth are signs of moderate to severe thalassemia, as the body produces more red blood cells to

make up for the anemia. Iron overload may result from the need for frequent blood transfusions in the most severe stage of the illness.

The most prevalent single-gene condition in the world is thalassemia. Thalassemia is particularly prevalent in the Mediterranean, the Middle East, Africa, South Asia, and Southeast Asia, where 2.5% to 25% of the population has the condition due to the partial protection against malaria that comes with inheriting the thalassemia gene. Although it is more common in these areas, thalassemia is present in persons from many different ethnic groups and geographical locations worldwide.

History

In 1925, thalassemia was initially identified in two forms: milder form (Cooley's anemia) and severe type. (Rietti-Greppi-Micheli's Malattia) The Thalassemia National Federation observed Thalassemia Day for the first time on May 8, 1994. The term "Mediterranean anemia" was coined since thalassemia condition was initially identified in people from Mediterranean nations (Europe, North Africa, and Southern Europe). It is estimated that 35 million people in India suffer from beta-thalassemia, a condition that is common throughout the nation. It is more common in particular racial groups, such as Parsis, Bengalis, Punjabis, and Gujaratis.

Classification of Thalassemia

1. Alpha thalassemia

When the alpha-globin gene is removed, the production of alpha-globin chains is decreased or ceases, resulting in alpha thalassemia. This gene has four alleles, and the number of them that are eliminated determines how severe the illness is. It is quite serious and can result in hydrops fetalis, which is lethal, if all four are removed. The condition is modest and frequently symptomless if only one is removed.

- **Hb Bart's Hydrops Foetalis**

The body stops making alpha-globin chains entirely when all four alpha-globin genes are eliminated (homozygous condition). This results in Hb Bart's hydrops fetalis, the most severe form of alpha-thalassemia. The four gamma-globin chains that make up Hb Bart's have a strong affinity for oxygen but are unable to release it into tissues, leading to severe tissue hypoxia.

- **HbH disease**

When three of the genes that produce hemoglobin's alpha chains are absent, hemoglobin-related hemoglobinopathy (HbH) results. As a result, the aberrant hemoglobin type known as HbH—which has four beta chains—is created. Hemoglobin's normal function is severely disrupted when alpha chains are absent. HbH aggregates inside the red blood cells to create formations known as Heinz bodies. The production of an abnormally long alpha chain results in Hb Constant Spring, a kind of HbH illness.

- **The α-thalassemia trait**

It can happen in two main ways:

The deletion of two of the four alpha-chain genes. This condition is known as homozygous α-thalassemia if it affects both copies of the gene. It is known as heterozygous α-thalassemia if it occurs in one copy of each gene (heterozygous); if only one alpha-chain gene is deleted, the condition is milder and is known as heterozygous α-thalassemia trait**.**

2. Beta thalassemia

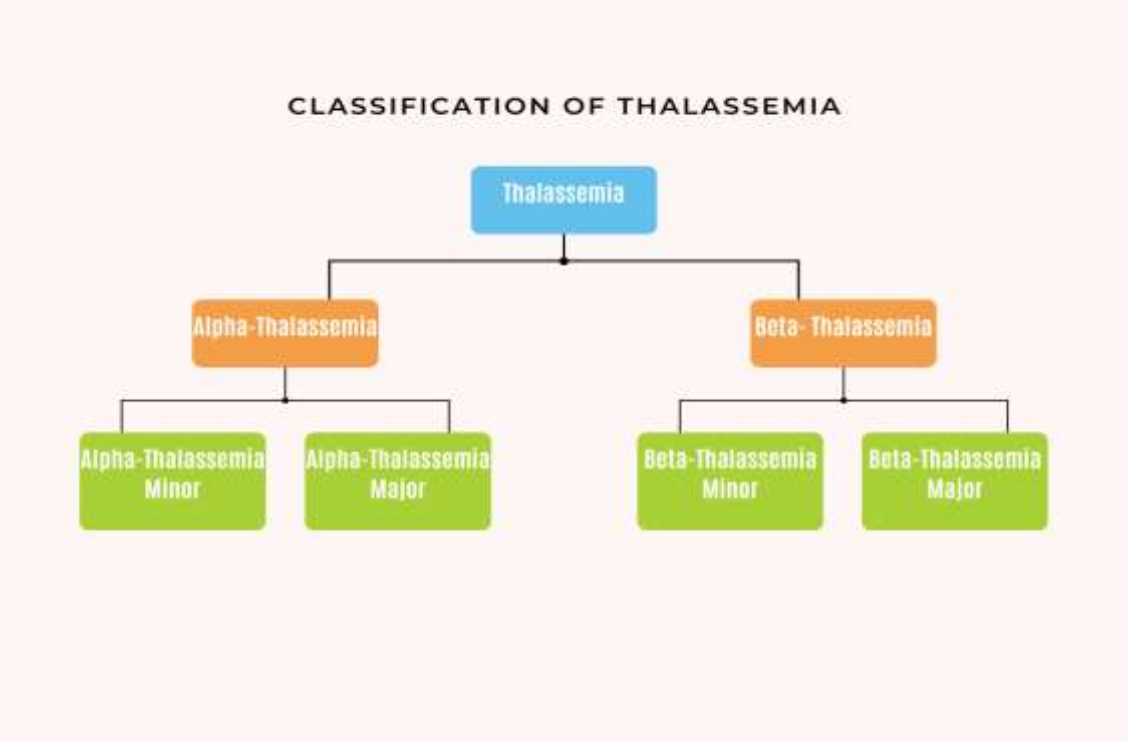
- Point mutations in the beta-globin gene cause beta thalassemia. Depending on the effect of the mutati-

on, it takes three forms:

- Beta-plus thalassemia: A single gene mutation results in minor symptoms that are typically undetectable.
- Both genes are defective in beta-thalassemia major, which causes severe symptoms like jaundice, sluggish growth, and frequent blood transfusions.
- The mild to severe symptoms of beta-thalassemia intermedia fall in between those of the other two forms.

Types
HB
HB-Electrophoresis
Genotype
Clinical Syndromes
3. Alpha-Thalassaemias
1. Hydrops foetalis
3-10 gm/dl
Hb Bart's (100%)
Deletion of four alpha- genes
Fatal in utero or in early infancy
2. Hb-H disease
2-12 gm/dl
HbF (10), HbH (2-4%)
Deletion of three alpha-gene
Haemolytic anaemia
3. Alpha- Thalassaemia trait
10-14 gm/dl
Almost normal
Deletion of two alpha-gene
Microcytic hypochromic blood picture but no anaemia
4. Beta-Thalassaemias
1. Beta-Thalassemia major
<5gm/dl
HbA(0-50%),
HbF(50-98%)
β-Thalassaemia
Severe congenital haemolytic anaemia, requires blood transfusions
2. Beta-Thalassemia intermedia
5-10 gm/dl
Variable
Multiple mechanisms
Severe anaemia , but regular blood transfusions not required
3. Beta-Thalassaemia minor
10-12 gm/dl
HbA (4-9%),

HbF (1-5%)
β/βThalassaemia
Usually asymptomatic



5. Pathophysiology

Although additional inherited genes may influence the severity of symptoms in each variety of thalassemia, the fundamental problem remains the same. Red blood cells (RBCs) have a shorter lifespan and hemoglobin (Hb) synthesis is reduced. This occurs as a result of the unaffected globin chains accumulating and harming the RBCs by forming unstable clumps inside them. RBC degradation and destruction occur early in beta-thalassemia because to the clumps' greater instability compared to alpha-thalassemia. Severe anemia and heightened RBC breakdown result from this.

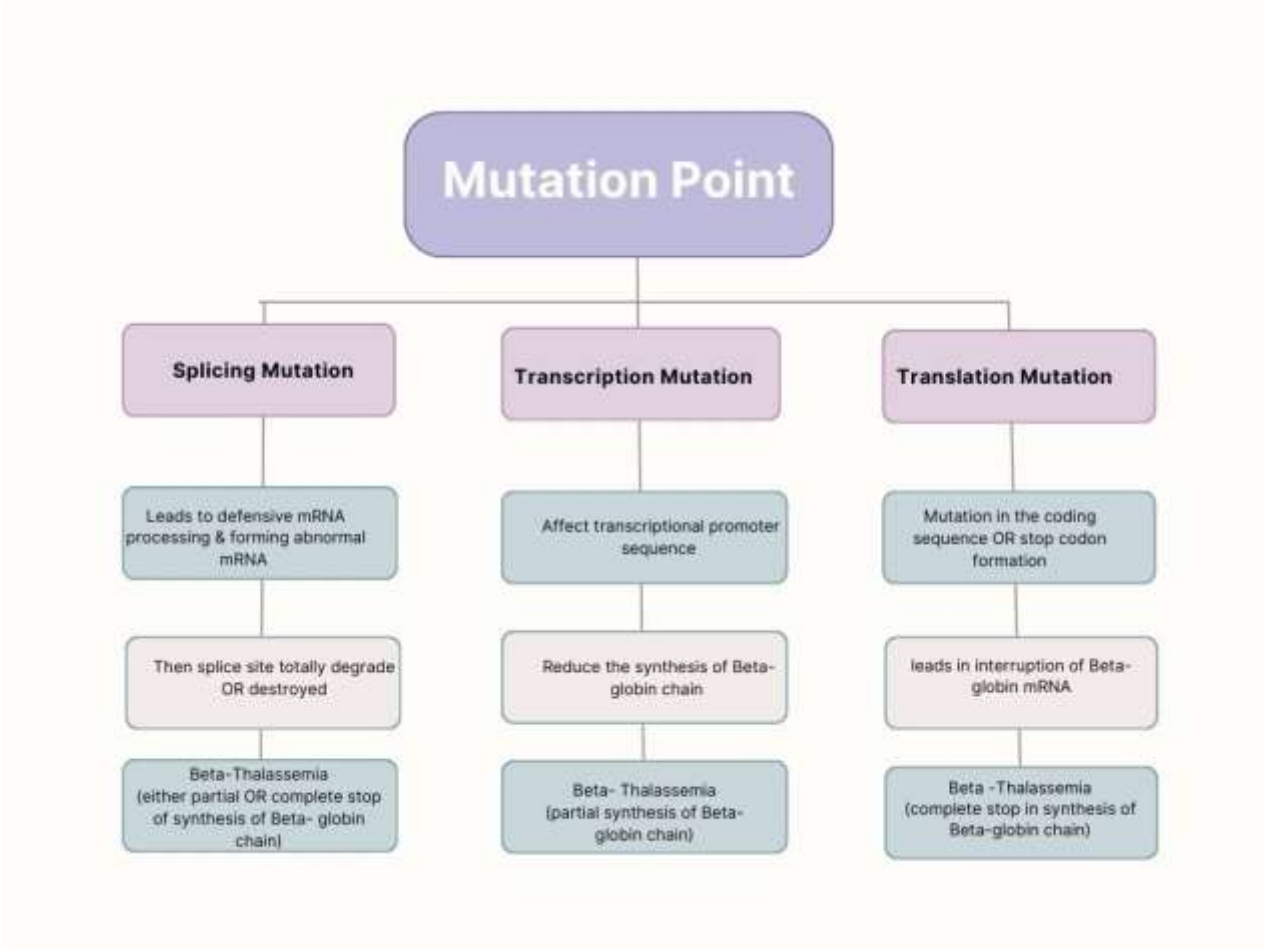
Bone abnormalities, particularly in the face and skull, can result from the body's attempt to compensate by creating more blood cells in the bone marrow in cases of severe beta-thalassemia. Furthermore, other organs such as the liver and spleen may produce more blood cells, which would result in their enlargement. In certain instances, it might also result in malignancies in these regions.

Alpha and beta-globin chains make up hemoglobin; a mutation in the beta-globin chain causes thalassemia. Splicing, transcription, and translation are the three RNA processes that can be impacted by mutation.

A biological process known as splicing mutation converts freshly generated pre-mRNA into mature mRNA.

The process by which cells create copies of RNA from a DNA fragment is known as a transcription mutation.

Using mRNA as a template, translation mutation is the process by which proteins are made in a cell.



- **Diagnosis**

When a regular blood test reveals mild anemia with smaller-than-normal red blood cells, people with thalassemia trait are frequently unintentionally identified. Other causes of this kind of anemia, known as microcytic anemia, include iron deficiency, lead poisoning, specific forms of anemia, and chronic illnesses. Doctors consider the patient's medical history, red blood cell size (MCV), and fluctuation in size (RDW) in order to determine the cause.

Except in cases of severe anemia, the MCV is rarely below 80 in iron deficiency and usually below 75 in thalassemia. The Mentzer Index, which is calculated by dividing the red blood cell count by the MCV, can be used to distinguish between thalassemia and iron insufficiency in children. Iron deficiency is indicated by a result above 13, whereas thalassemia is indicated by a score below 13.

Because the RDW is frequently larger in iron deficiency and lower in thalassemia, it can also be used to distinguish between the two conditions. Thalassemia is more likely if RDW is normal, but more testing is required if RDW is excessive.

A blood smear, hemoglobin electrophoresis (a test for various forms of hemoglobin), ferritin levels (a good indicator of iron deficiency), lead levels, and, in rare cases, bone marrow examination are further procedures. The most reliable indicator of iron deficit is ferritin; if it is normal, iron insufficiency is improbable. Lead poisoning can be ruled out with a normal lead level.

Hemoglobin electrophoresis is frequently performed to confirm thalassemia if it is suspected. The test typically reveals higher levels of HbA2 and HbF and lower levels of HbA in beta thalassemia. HbA2 levels, however, may seem normal if the patient also has iron insufficiency, which may complicate the

diagnosis. Certain hemoglobin types (Hb Bart's or HbH) in neonates may be indicative of alpha thalassemia.

6. Case Study:

- **Case study1**

This is a thorough case study of a 17-year-old kid from West Bengal, India, who had a variety of symptoms, such as exertional dyspnea, weariness, jaundice, and abdominal pain. Congenital hemolytic anemia caused by compound heterozygosity for HbE/Beta Thalassemia was the diagnosis.

- **Presenting Complaints:**

a. Fatigue for 8 years
b. Unusual size of head for 6 years
c. Yellowish discoloration of eyes for 8-9 month
d. Abdominal discomfort and exertional dyspnea for 8 months

- **Family History:**

One brother has a similar illness but not severe and not taking any treatment

- **Physical Examination:**

1. Severe pallor
2. Mild icterus
3. Frontal bossing
4. Flat nasal bridge
5. Liver palpable 3 cm below RCM with a liver span of 15 cm
6. Massive splenomegaly

- **Investigations:**

1. CBC: TC-5600, DC- N76 L12, Hb-5.6, HCT-16.4, MCV-68, MCH-20.9, MCHC-30, RDW- 30.5, PLT- 4.8L
2. Reticulocyte count: 5.8%
3. S. Ferritin: 315 (23-322)
4. Osmotic fragility test: negative
5. P. Smear: Microcytic hypochromic anemia, Microspherocytes, and fragmented RBCs with evidence of hemolysis
6. HPLC: Hb A2- 67% and Hb F-29%
7. Hb electrophoresis: Hb fractions Hb A-6.7%, Hb F- 34.2%, Hb E- 53.2%, Hb A2- 5.9%

- **Diagnosis**:- Congenital hemolytic anemia due to compound heterozygosity for HbE/Beta Thalassemia
- **Treatment:-** Packed cell transfusions to raise the Hb

This instance emphasizes the significance of thalassemia early detection and treatment. Thalassemia is a hereditary condition that, if untreated, can cause severe anemia and other consequences.

- **Case Report 2**

A 5.5-year-old Maldivian girl with the inherited blood condition Beta Thalassemia Major is the subject of this case study. Severe anemia results from beta thalassemia's decreased hemoglobin synthesis. Due to the high incidence of malaria, which raises the number of instances of Thalassemia, this condition is widespread in places like the Maldives.

Although the girl was initially diagnosed with beta thalassemia at the age of two, she did not receive regular treatment follow-up. She later came back to the hospital with symptoms like fatigue, irritation,

shortness of breath, and coughing. A physical examination showed that she was extremely anemic, had pale skin, fragile hair, and nails, and had developed "chipmunk facies," a characteristic of untreated beta thalassemia.(a facial abnormality brought on by bone growth)

Her hemoglobin level (4.5 gm/dl) was extremely low. She has Beta Thalassemia Major, according to a blood test. After receiving a blood transfusion without any problems, her hemoglobin level increased to 9.5 gm/dl. However, she also had high amounts of iron in her body (ferritin level of 3562.69 ng/ml) as a result of her frequent need for blood transfusions. To get rid of the extra iron in her body, she needed to be treated with iron-chelating medications.

In addition to routine testing to check on her liver, thyroid, and general growth, the girl now needs blood transfusions every 20 days**.**

- **Case Report 3**

On January 23, 2020, an 8-year-old boy from Shirajgam was brought to AVBRH's Pediatric Ward No. 14 with symptoms of fever, low hemoglobin levels, and abdominal pain. At the age of eight months, he was diagnosed with thalassemia major. He was examined in a number of ways upon arrival, and treatment was started right away. After beginning treatment, he demonstrated notable progress, which persisted until the day I left. Preventive interventions are advised because thalassemia is one of the most prevalent hereditary disorders in the world and necessitates long-term care. For carriers, screening and counseling are typically voluntary.

To assess the clinical presentation and treatment of thalassemia, a study called "A Clinical-Epidemiological Study of Thalassemia Cases in India" was conducted. For this investigation, patient records from 2005 to 2014 were examined. Out of the 183 instances, 179 (97.8%) had beta-thalassemia major, 3 (1.6%) had beta-thalassemia intermedia, and 1 (0.6%) had beta-thalassemia minor, according to the results. A quarter of cases were diagnosed during the first six months, and the majority were diagnosed by the age of one. Thirteen patients (7.1%) had bone deformities, 172 patients (94%), 179 patients (97.8%) had pale skin, and 34 patients (18.6%) had fever. More than half of children under five were found to be stunted, and one-third were underweight.

After receiving transfusions for a year, the average hemoglobin level was 10 ± 1.6 g/dL. Starting at an average age of 11.1 years, 51 patients (27.9%) were treated with desferrioxamine as part of iron chelation treatment. At an average age of 10.7 years, the spleen was removed in 4 cases. A higher incidence of lenticular opacity was linked to the usage of desferrioxamine. According to the study's findings, thalassemia patients face a number of consequences, underscoring the necessity of specialist care to adequately address these problems.

- **Treatment:**

Beta thalassemia major includes following Treatments:

- **Blood transfusions:** A patient with mild or severe thalassemia will receive blood intravenously during this treatment. The frequency of blood transfusions will depend on the type of thalassemia. The process can take one to four hours. Blood transfusions are frequently necessary for more severe types, perhaps every three to four weeks. To maintain a healthy supply of red blood cells in the blood, repeated transfusions are necessary. Sometimes blood transfusions are necessary for people with hemoglobin H or beta-thalassemia intermedia. Coolley's anemia, also known as beta-thalassemia major, requires frequent blood transfusions every two to four weeks.
- **During Iron Chelation Treatment**

RCB is a protein high in iron. Iron overload is a blood disorder caused by frequent blood transfusions that

accumulate iron. It damages the heart, liver, and a few other bodily components. The purpose of iron chelation therapy is to eliminate excess iron from the body and avoid damage. Two medications are utilized in this treatment:

Injections of deferoxamine are used to remove excess iron from the body in patients with thalassemia or anemia.

For individuals with non-transfusion-dependent Thalassemia, deferasirox, an oral iron chillator, is used to treat chronic iron overload. (NTDA)

- **Folic Acid Supplements**

Folic acid is vitamin B which helps in increasing low level of folic acid in RBC.

- **Blood and Marrow Stem Cell Transplant**

Stem cells and blood The purpose of a transplant is to replace damaged stem cells with healthy ones from a healthy donor.

- **Other Treatment Includes:**

Allogenic Haemopoietic Cell Transplantation
Ery Thyroid Maturating Agents
Gene Therapy: Gene Insertion Approaches

- **Future Treatments:**

Gene Therapy: Gene Edition Approaches
Iron Metabolism Modifying Agents
(Minihepcidines, Ferroportion Inhibitors.

- **Perventions**

To better manage these conditions, we need to focus on several key actions:

- **Education and Awareness:** Raise public, student, and health professional knowledge and understanding of these illnesses.
- **Screening and Diagnosis:** Establish extensive screening programs and provide facilities for prenatal diagnosis and genetic counseling, particularly in remote areas.
- **Patient Care:** To continue to handle patients with thalassemia, more day care centers should be established.
- **Affordable Treatment:** Create more affordable stem cell transplant options.

- **Conclusion**

The Impact of Thalassemia on Social Life

Thalassemia, a genetic blood disorder, has a profound impact on the social lives of individuals affected by it. The chronic nature of the disease, coupled with the need for regular blood transfusions and medical check-ups, can lead to significant social and emotional challenges.

The effects of thalassemia on social life can be far-reaching, influencing relationships, education, career choices, and overall well-being. Individuals with thalassemia may experience feelings of isolation, stigma, and anxiety, which can further exacerbate the physical symptoms of the disease.

Key findings of the impact of thalassemia on social life include:

1. **Social isolation:** Thalassemia can lead to social isolation, as individuals may avoid social interactions due to feelings of embarrassment or shame about their condition.
2. **Stigma and discrimination:** Thalassemia can be stigmatized, leading to discrimination and social exclusion.

3. **Emotional distress:** The chronic nature of thalassemia can lead to emotional distress, including anxiety, depression, and stress.
4. **Impact on education and career:** Thalassemia can impact educational and career choices, as individuals may need to prioritize their health over other aspects of their life.
5. **Strained relationships:** Thalassemia can strain relationships with family and friends, who may not fully understand the condition or its impact on daily life.

In conclusion, thalassemia has a significant impact on the social lives of individuals affected by it. It is essential to raise awareness about thalassemia and its effects on social life, to promote understanding, acceptance, and support for individuals with the condition. Additionally, healthcare providers, educators, and employers must work together to provide comprehensive support and accommodations to help individuals with thalassemia lead fulfilling and inclusive lives.

Quality of Life in Thalassemia :

Modern treatments have significantly reduced the number of deaths and illnesses associated with thalassemia. Now, enhancing patients' quality of life (QOL) should also be a priority. Because QOL examines several aspects of a person's life and takes into account their own viewpoint on their well-being, it provides a more comprehensive picture than standard medical evaluations. On the other hand, little study has been done on how to assess QOL in thalassemia patients.

Reliability, sensitivity to the main difficulties faced by thalassemia patients, and adaptability to various cultures, ages, and social contexts are all important characteristics of a solid QOL measurement tool. Comparing health outcomes between clinics and assessing novel treatments would both benefit from this type of technology. Two instruments that measure the emotional and medical difficulties that patients, parents, and kids confront have been tested: one is based on the WHOQOL-100 questionnaire, while the other is designed especially for thalassemia. To develop and evaluate such technologies for thalassemia patients, more study is required.

Asking patients directly about their regular treatment and how it impacts their quality of life is another strategy. According to surveys conducted in the UK and Cyprus, patients expressed dissatisfaction with the way blood transfusions are handled, a dearth of chelation therapy choices, and inadequate communication with medical professionals. These are issues of practicality that could be made better.

Social Consequences of Thalassemia:

Thalassemia puts a lot of stress on both the patient and their family. Modern treatments have improved life expectancy for people with the disease, so it is now even more important to address the emotional and social challenges they face.

This study aimed to understand the emotional and social struggles of teenagers with thalassemia. Interviews were conducted with 50 teenagers, and their experiences were studied in areas like education, sports, family life, and social interactions.

Education: Many teenagers felt that thalassemia had a negative effect on their school performance. Previous studies, such as one conducted in the UK, showed that most students with thalassemia had to miss school due to their illness, which matched the findings of this study.

Sports: Information on how thalassemia affects sports is limited, but many teenagers in this study reported feeling tired and unable to perform well in sports, which aligns with other research.

Self-esteem and appearance: Thalassemia can cause physical changes like delayed growth, short height,

and changes to facial features due to the disease. These changes often make teenagers feel different, leading to low self-esteem and fear of rejection because of their appearance.

Family relationships: Unlike studies in other countries where patients felt disconnected from their families, the teenagers in this study felt that their family relationships were strong. They often shared their worries with their parents, possibly because Indian families tend to be close-knit and offer support for a longer time.

Anxiety and mental health: Many teenagers with thalassemia feel anxious, especially those who experience physical changes due to the disease. Studies have shown that patients often have low self-esteem and are worried about pain, death, and the unknown, similar to what was found in this study.

Reference

1. Giardina P, Forget B. Thalassemia syndromes. In:Hoffman R, Benz E, Shattil S, et al, eds. Hematology: Basic Principles and Practice (5th ed). Phila-delphia, PA: Churchill Livingstone; 2008:535-563.
2. Weatherall DJ. Genetic variation and susceptibility to infection: the red cell and malaria. Br J.Haematol. 2008;141(3):276-286.
3. Weatherall DJ. The inherited diseases of hemoglobin are an emerging global health burden. Blood. 2010;115(22):4331-4336.
4. Weatherall D. The thalassemias. In:Stamatoyannopoulos G, Nienhuis A, Majerus Peds. Molecular Basis of Blood Diseases (2nd ed).Philadelphia, PA: Saunders; 1994:157.
5. Rund D, Rachmilewitz E. Beta-thalassemia.N Engl J Med. 2005;353(11):1135-1146.
6. Karimi M, Haghpanah S, Farhadi A, Yavarian M. Genotype-phenotype relationship of patients with beta-thalassemia taking hydroxyurea: a 13-year experience in Iran. Int J Hematol 2012;95:51e6.
7. Banan M, Bayat H, Azarkeivan A, Mohammadparast S,Kamali K, Farashi S, et al. The XmnI and BCL11A single nucleotide polymorphisms may help predict hydroxyurea response in Iranian beta-thalassemia patients. Hemoglobin 2012;36:371e80.
8. Kohne E. Hemoglobinopathies: clinical manifestations, diagnosis, and treatment. Dtsch Arztebl Int2011;108:532e40.
9. Castro O, Nouraie M, Oneal P. Hydroxycarbamide treatment in sickle cell disease: estimates of possible leukaemia risk and of hospitalization survival benefit. Br J Haematol 2014;167:687e91.
10. Sankaran VG, Menne TF, Xu J, Akie TE, Lettre G, Van Handel B, et al. Human fetal hemoglobin expression is regulated by the developmental stage-specific repressor BCL11A. Science 2008;322:1839e42.
11. Sedgewick AE, Timofeev N, Sebastiani P, So JC, Ma ES, Chan LC, et al. BCL11A is a major HbF quantitative trait locus in three different populations with beta-hemoglobinopathies. Blood Cells Mol Dis 2008;41:255e8
12. Payen, E., and Leboulch, P. (2012). Advances in Stem Cell Transplantation and Gene Therapy in the β-hemoglobinopathies. Hematol. Am Soc Hematol Educ Program 2012, 276–283. doi:10.1182/asheducation-2012.1.276
13. Persons, D. A., Allay, E. R., Sabatino, D. E., Kelly, P., Bodine, D. M., and Nienhuis, A. W. (2001). Functional Requirements for Phenotypic Correction of Murine Beta-Thalassemia: Implications for Human Gene Therapy. Blood 97 (10), 3275–3282. doi:10.1182/blood.v97.10.3275

14. Psatha, N., Karponi, G., and Yannaki, E. (2016). Optimizing Autologous Cell Grafts to Improve Stem Cell Gene Therapy. Exp. Hematol. 44 (7), 528–539. doi:10.1016/j.exphem.2016.04.007
15. Puthenveetil, G., Scholes, J., Carbonell, D., Qureshi, N., Xia, P., Zeng, L., et al. (2004). Successful Correction of the Human Beta-Thalassemia Major Phenotype Using a Lentiviral Vector. Blood 104 (12), 3445–3453. doi:10.1182/blood-2004-04-1427
16. Rachmilewitz, E. A., and Giardina, P. J. (2011). How I Treat Thalassemia. Blood 118 (13), 3479–3488. doi:10.1182/blood-2010-08-30033
17. Ross and Wilson anatomy and physiology in health and illness , 12th edition anne Waugh allison grant.
18. Mohan Hasrh ,Text book of Pathology 8th edition Page no 335 to 339
19. Ishfaq K, Naeem SB, Ahmad T, Zainab S, Psychosocial and economic impact of Thalassemia Major on patient families. Isra Medical Journal 2016 Jan 1(8)
20. Ratip S, Modell B. Psychological and sociological aspects of the thalassaemias. Seminars in Hematology 1996; 33: 53-65.
21. Tsiantis J, Xypolita-Tsantili D, Papadakou-Lagoyianni S.Family reactions and their management in a parents group with b-thalassaemia. Archiv Dis Child 1982; 57: 860-863.
22. Logithis J, Haritos-Fatouros M, Coonstantoulakis M.Intelligence and behavioral paterns in patients with Cooley's anaemia; a study based on 138 consecutive cases. Pediatr 1971;48: 740-743.
23. Ratip S, Skuse D, Porter J. Psychosocial and clinical burden of thealassaemia intermedia and its implications for prenatal diagnosis. Arch Dis Child 1995; 72: 408-412.
24. Georganda ET. The impact of thalassaemia on body image, self-image and self-esteem. Annals New York Academy of Sciences 1998; 850: 466-472.
25. Sherman M, Koch D, Giardiana P. Thalassaemic children's understanding of illness: A study of cognitive and emotional factors. Annals New York Academy of Sciences 1985; 445: 327-336.
26. Woo R, Giardina P, Hilgartner M. A psychosocial needs assessment of patients with homozygous ~-thalassemia. AnnNY Acad Sci 1985; 445: 316-323.
27. Massaglia P, Carpignano M. Psychology of the thalassaemia patient and his family. Thalassaemia today; The Mediterranean experience. Proceedings of the 2 "d Mediterranean Meeting, Milano. 1985: 28-30.