DOI: https://doi.org/10.53350/pjmhs2023176257

ORIGINAL ARTICLE

Efficacy of Education in Providing Knowledge about Thalassemia Prevention and Family Screening of Thalassemic Patients

RAMEEZ IQBAL[1], KIRAN MEMON[2], SURESH KUMAR[3], MUHAMMAD RAHIL KHAN[4], SHABNAM[5], AMANULLAH BHUTTO[6], KIRAN AAMIR[7], AAMIR RAMZAN[8]

[1]*Lecturer,Pathology Department Liaquat university of Medical and health sciences jamshoro Hyderabad*
[2]*Assistant Professor pathology Department,Indus medical college,Tando Muhammad khan*
[3]*Assistant Professor, Pathology department Jinnah sindh medical university Karachi*
[4]*Assistant professor,Pathology Department,Liaquat university of Medical and health sciences jamshoro Hyderabad*
[5]*Lecturer ,Pathology Department Liaquat university of medical and health sciences jamshoro Hyderabad.*
[6]*Assistant Professor,Pathology Department,Ghulam Mohammad Mahar Medical College Sukkur*
[7]*Associate professor,Pathology Department,Liaquat university of Medical and health sciences jamshoro Hyderabad*
[8]*Lecturer in pathology Department, Liaquat university of Medical and health sciences jamshoro Hyderabad*
Corresponding author: Rameez Iqbal, Email: rameezmemon30@yahoo.com, Cell: 03322056030

ABSTRACT

Objective: The purpose of this study was to track out Beta Thalassemia carriers in affected households and evaluate the efficacy of thalassemia awareness campaigns.

Study Design: Observational - Cross-Sectional study

Place and Duration: Department of Pathology & Diagnostic and Research Laboratory - LUMHS Hyderabad/Jamshoro. 01-09-2021 to 31-03-2022

Methods: A total of 55 thalassemia patients and families included in the study after providing written consent; the research was conducted at the Department of Pathology and Diagnostic and Research Laboratory. Demographic information such as age, gender, education level, and family history of thalassemia was recorded, and SPSS 24.0 was used for statistical analysis.

Results: In 55 patients, majority 33 (60%) were males and 22 (40%) were females. The frequency of bimonthly blood transfusions was found to be the lowest, and consanguineous marriage between parents was found in 46 (83.6%) of patients. Three (5.5%) siblings of thalassemia patients were found to have thalassemia major, and seventeen (34.5%) siblings were found to have thalassemia minor. Thirty-five (63.6% of patients) were found to have a normal electrophoresis pattern.

Conclusion: The research showed that roughly 50% of families were impacted by thalassemia and that 25% of individuals had previously received a blood transfusion. The instructional event was evaluated positively for its impact on participants' understanding of thalassemia.

Keywords: Effectiveness of Educational Awareness, Carriers, Thalassemia, family history

INTRODUCTION

Population screening is an established practice that has been refined since its inception in the 1960s as new information about illness development and prognosis has become available[1]. Given the high mortality rate among thalassemia patients, public health innovation guidelines recommend using premarital screenings to better ensure healthy offspring. Treatments for thalassemia major place a substantial financial burden on public health budgets[2] because to the disease's negative effects on patients' and their families' health, social, and economic well-being.

A decrease or absence of hemoglobin chains arises in thalassemia patients because of a disruption in the amino acids that make one or more globin chains in red blood cell hemoglobin[3]. The prevalence of thalassemia in Indonesia makes it the country's most prevalent genetic condition. The prevalence of beta-thalassemia gene carriers is estimated to be 3-5% globally, and as high as 10% in some regions[4]. In Indonesia, the frequency of thalassemia is highest in the province of West Java, where as much as 42% of the total population of about 6647 individuals are affected.4. Premarital screening for thalassemia is essential since patients with the disease need regular blood transfusions and iron chelation for the rest of their lives. Preventing thalassemia is seen as beneficial to society and financially as well[5].

Thalassemia is a genetic condition with a wide range of genetic defects that cause varying degrees of globin chain synthesis impairment, leading to a wide range of clinical presentations. Countries like Italy, Greece, and Cyprus have shown that this disease is preventable. They pioneered national programs that have significantly reduced the number of affected births, and the World Health Organization has warned that without such measures, most countries will be unable to provide optimal treatment to all thalassemic patients due to the high cost of doing so. The cost of prevention in Iran remains stable each year, while the cost of treatment continues to climb. Mazandaran Province's National Thalassemia Prevention Program in Iran proved that a catastrophic loss of income can be avoided.[6]Similar good effects of similar training programs on parents' disease awareness have been observed in other research.[7] Main prevention measures include screening and counseling of high-risk families, screening of the general population before marriage, and dissemination of accurate information to the public and professionals.[8] Prenatal diagnosis and counseling for at-risk couples, as well as adequate and extensive screening, are viable strategies for reducing thalassemia-related mortality and morbidity in areas where the disease is endemic.[9] Because to premarital screening, the incidence of -thalassemia major in newborns has dropped dramatically.[10,11]

Couples at risk of having problems in their marriage or having children may benefit from being screened before they get married. However, in traditional Asian societies, marriage is a highly intricate business. If both partners are already committed, a high-risk couple may decide to go through with the wedding anyhow. Breaking off a marriage is often seen with shame and disgrace from friends and family [11]. A research conducted in Saudi Arabia confirmed these findings; after obtaining genetic counseling, over 90% of high-risk couples still went forward with the marriage. Possible contributing factors to this result in Saudi Arabia include the lack of premarital screening and a lack of education about the risks associated with having thalassemic children. In fact, despite the implementation of premarital screening in Saudi Arabia [12], a recent study indicated that over half of the college students sampled had never heard of thalassaemia. Therefore, education is crucial to the success of any program aimed at reducing the incidence of thalassemia. Understanding the cultural context is essential to any effort to increase community awareness, as it is the source from which people derive their beliefs, attitudes, and perceptions. According to the results of these studies, a thalassemia preventive program will have less of an impact if there isn't widespread public awareness of it.

Approximately 72% of Bangladesh's roughly 160 million people reside in rural areas, making it a low-income country. Bangladesh falls inside the global thalassaemia belt, but little is known about the disease's epidemiology or natural history. Bangladesh lacks structured national programs for thalassemia awareness, carrier screening, or patient management [14,15], in contrast to its South Asian neighbors. PND is not commonly available in Bangladesh, and the social acceptability of pregnancy termination is unclear. Furthermore, there is likely to be widespread ignorance and false beliefs regarding thalassaemia.

Multicenter research found that 26% of parents of children with beta-thalassemia major and 61% of parents overall were illiterate; however, no parents had undergone prenatal screening and just 10 had undergone premarital screening [11]. Consanguineous marriages are prevalent in today's culture, despite the lack of premarital thalassemia test or counseling. There is a lack of access to antenatal care, and termination of pregnancies raises moral and theological questions. Since bone marrow transplantation is not an option for such a high number of individuals with beta-thalassemia major, the disease is controlled with blood transfusions, which causes problems and ultimately death. If the mother's life is in jeopardy or there are severe abnormalities in the fetus, an abortion is permitted in Islam up to 120 days into the pregnancy [10].

There is now no medication or treatment that may cure this genetic mutation condition, but you can take steps to protect yourself by not getting married to someone who is a carrier and getting a thalassemia blood test regularly. There are many reasons why thalassemia screening should be widely publicized, including the disease's high incidence rate, high severity, and significant societal impact.

MATERIALS AND METHODS

The Observational - Cross-Sectional study was conducted at the Department of Pathology & Diagnostic and Research Laboratory - LUMHS Hyderabad/Jamshoro on a sample of 55 patients. All patients, both male and female, with a confirmed diagnosis of b-thalassemia were taken into account. Individuals with different hemoglobinopathies or who did not provide written consent were not included.

A blood sample was taken from each subject. Following standard precautions, a blood sample of 3 mL will be drawn using a 5 cc disposable syringe and deposited in a purple-capped tube containing ethylene diamine tetra acetic acid (EDTA). To acquire an accurate blood count, a Japanese fully-automatic analyzer, the Sysmex XN1000, will be employed. The electrophoresis will be carried out using a High Performance Liquid Chromatography (HPLC) instrument.

The EDTA-treated samples were run through an automated hematology analyzer from Sysmex XN1000i Japan to get a full blood count.

The data was analyzed using SPSS 24.0 and Microsoft Excel 2016. There were no &% indicators to indicate quality. A mean and standard deviation (X SD) formula was used to represent numerical data. The Chi-Square test was used to compare students' levels of knowledge acquisition before and after the tutorial. A p-value less than 0.05 will be considered statistically significant.

RESULTS

In 55 patients, majority 33 (60%) were males and 22 (40%) were females.(Figure 1)

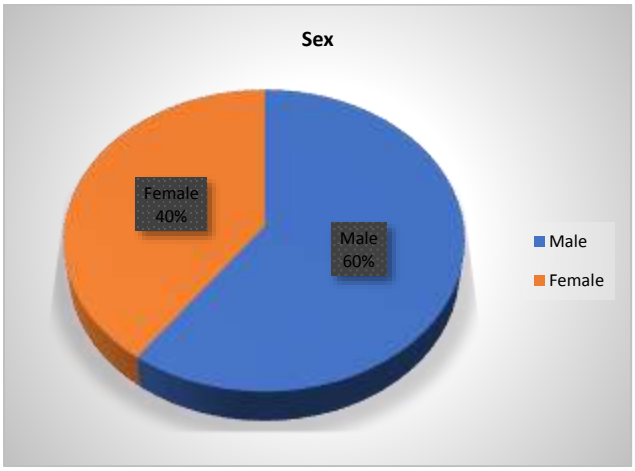

Figure 1: Sex distribution of all cases

Twenty-two patients (40 percent) had transfusions every fortnightly, 19 patients (34.5 percent) receiving transfusions every month. The frequency of transfusions reported as bimonthly was lowest.(table 1)

Table-1: Blood transfusion and its frequency

Variables	Frequency (55)	Percentage
Blood Transfusions		
fortnightly	22	40
Weekly	5	9.1
Twice a week	6	10.9
Monthly	19	34.5
Bimonthly	3	5.4

Consanguineous marriage between parents was found in 46 (83.6%) of patients.(table 2)

Table 2: Marriage details of patients

Variables	Frequency (55)	Percentage
Consanguineous marriage		
Yes	46	83.6
No	9	16.4

Three (5.5%) siblings of thalassemia patients were found to have thalassemia major, and seventeen (34.5%) siblings were found to have thalassemia minor.(table 3)

Table 3: Severity of thalassemia

Variables	Frequency (55)	Percentage
Thalassemia		
Major	3	5.5
Minor	17	34.5
Normal	25	45.4

Table 4: We found significantly improvement in knowledge after providing education about thalassemia. (table 4)

Statements	Appropriateness of Knowledge				P - Value
	Pre-Test – Before Educational Session		Post-Test – After Educational Session		
General Knowledge about Thalassaemia:					
Thalassemia is a Blood Disease	97.2%	2.8%	71 97.2%	2.8%	> 0.05
Thalassemia is more common in males	28.76%	71.24%	4.1%	95.9%	> 0.05
Thalassemia is more common in females	71.24%	28.76%	5.48%	94.52%	> 0.05
Thalassemia is equally present in both gender	27.4%	72.6%	91.8%	8.2%	< 0.05*
Thalassemia can be diagnosed via Blood Tests	91.8%	8.2%	100%	0 (0%)	< 0.05*
Knowledge about Diagnosis of Thalassaemia					
Thalassemia can only be diagnosed after birth of a child	97.2%	2.8%	19.1%	80.8%	< 0.05*

Thalassemia can be diagnosed before birth of a child	9.6%	90.4%	100%	0 (0%)	> 0.05
Knowledge about transmission of Thalassaemia					
Thalassemia can be transmitted from Parents to Offspring	86.3%	13.7%	100%	0 (0%)	> 0.05
Chances of having thalassemia are increased if there is cousin marriage among parents	43.9%	56.1%	100%	0 (0%)	< 0.05*
Marriage between 2 thalassemia carrier leads to a thalassaemic child	67%	33%	100%	0 (0%)	< 0.05*
Marriage between healthy and a carrier leads to thalassaemic child	58.9%	41.1%	2.8%)	97.2%	> 0.05
Knowledge about prevention of Thalassaemia					
Thalassaemia is a preventable disease	21.9%	78.1%	100%)	0 (0%)	< 0.05*
If a family has history of thalassemia, would there be any consanguineous marriage be done	69.9%	30.1%)	21.9%	78.1%	> 0.05
Thalassemia carriers should not marry	21.9%	78.1%	71.2%	28.8%	> 0.05
Thalassemia carrier couple should not have pregnancy	50.7%	49.3%	86.3%)	13.7%	< 0.05*
Thalassemia carriers should have pre-marital screening	63%	37%	100%	0 (0%)	> 0.05

DISCUSSION

Thalassemia is an inherited disorder that disrupts the body's ability to produce healthy red blood cells. [16] Complications from the blood's hazardous excess of iron necessitate lifelong therapy, including regular blood transfusions and the administration of iron chelation. Taking precautions before testing for a genetic condition is crucial. Prenatal genetic testing is strongly recommended for all couples carrying thalassemia genes. Awareness of genetic screening needs a solid foundation in thalassemia knowledge.[17]

According to the results, the vast majority of respondents lacked basic information. Knowledge among adolescents has been studied in a variety of ways. The same was true of an Iranian study; just around 20% of students there have a solid foundational knowledge of thalassemia. However, other research out of India reveals significantly better outcomes, with the vast majority of respondents grasping the concepts easily. [18]

Research has consistently shown that males and females learn about thalassemia differently. According to studies conducted in Iran, there are much more female students than male students with a deep understanding of thalassemia. Basu's study, on the other hand, found that male respondents tend to have more knowledge than their female counterparts. [19] However, there is no consideration for disparities between the sexes in this analysis.

Respondents have an adequate foundational understanding of thalassemia's pathogenesis, symptoms, and treatment. Respondents also have a limited grasp of the concepts of definition, etiology, onset, and prevention. Most respondents believe microorganisms and unhealthy lifestyles can cause thalassemia, hence it is important to highlight the topic of aetiology while promoting health in relation to the disease. Respondents' knowledge of thalassemia prevention is hampered by their ignorance of the significance of genetics in the disease's causation. Some of those polled had no idea that thalassemia prevention efforts included genetic screening. The vast majority of respondents have maintained a diet and healthy lifestyle that can help stop the spread of thalassemia. [20]

Respondents also show a lack of comprehension regarding the beginning of thalassemia. Most people who participated in the survey believed that thalassemia was present at birth, while more than half of those who participated incorrectly believed that previously healthy people may get the disease. Most respondents are aware that blood transfusion is the treatment for thalassemia, however a sizable minority of them selected "dialysis" as their option. End-stage renal failure is typically treated with dialysis or hemodialysis, however this is unrelated to thalassemia. [21]

Nearly a third of respondents admitted they were familiar with thalassemia, but the vast majority were not. When compared to previous studies conducted in a variety of nations, a substantially smaller proportion of respondents in this study reported prior knowledge of thalassemia. According to the findings of Ebrahim et al., 53 percent of respondents reported previously learning about thalassemia. Balcin found that 57.7% of respondents were familiar with thalassemia, whereas study in India found an even greater figure, as high as 85% of respondents. [22]

This study's findings of poor thalassemia awareness highlight the disease's considerable prevalence despite its lack of public attention. The incidence of thalassemia is very high in Indonesia and other nations in the "thalassemia belt." Thalassemia is a genetic blood disorder that requires lifelong care, which can be emotionally and financially taxing on families and nations. The number of thalassemia instances in a country can be decreased by the implementation of preventative programs, such as genetic counseling. [23] Public education on thalassemia as a genetic disease, along with public education about genetic counseling (33%), and public education about premarital screening (27%), can have a significant impact on the success of the thalassemia preventive program, which accounts for up to 40% of the whole program. [24]

People who carry the thalassemia gene (carrier thalassemia) may go unnoticed since they may not show the same severe symptoms as those who have thalassemia major. The fact that thalassemia carriers often have anemia is well-known. [25] The first stage in raising public awareness of thalassemia is educating the public, especially kids who will be screened for the thalassemia gene, on the disease's severity, causes, and symptoms. Respondents' preferred means of learning about thalassemia were the internet, classroom instruction, health professionals, seminars, family and friends, and the print media. A component of thalassemia preventive programs can be found in high school health programs, which typically involve frequent counseling and lectures open to students of all disciplines. Both traditional methods of delivery, such as lectures, and interactive methods, such as group discussions, can be used to impart knowledge at seminars. When compared to traditional lecture approaches, which only result in a 20% improvement in student understanding, group discussion is far more effective. Because education is crucial to raising awareness of thalassemia, preventative programs aimed at teenagers should begin as soon as possible. [26]

CONCLUSION

The research showed that roughly 50% of families were impacted by thalassemia and that 25% of individuals had previously received a blood transfusion. The instructional event was evaluated positively for its impact on participants' understanding of thalassemia.

REFERENCES

1. Gason, A.A., M.A. Aitken, S.A. Metcalfe, K.J. Allen and M.B. Delatycki 2005. Genetic susceptibility screening in schools: attitudes of the school community towards hereditary haemochromatosis. Clin. Genet., 67: 166-174.
2. Weatherall, D.J., 2008. Genetic variation and susceptibility to infection: The red cell and malaria. Br. J. Haematol., 141: 276-286.
3. Galanello, R. and R. Origa, 2010. Beta-thalassemia. Orphanet J. Rare Dis., Vol. 5.
4. Yayasan Thalassemia Indonesia, 2017. Program pencegahan penyakit thalassemia. YTI-POPTI Jabar, Bandung, pp: 2
5. Ahmadnezhad, E., N. Sepehrvand, F.F. Jahani, S. Hatami, C. Kargar, M. Mirmohammadkhani and S. Bazargan-Hejazi, 2012. Evaluation and cost analysis of national health policy of thalassaemia screening in West-Azerbaijan province of Iran. Int. J. Prev. Med., 3: 687-692.
6. Karimi M, Johari S, Cohan N. Attitude toward prenatal diagnosis for β-thalassemia major and medical abortion in Southern Iran. Hemoglobin 2010; 34:49-54.

7. Dehkordi AH, Heydernejad MS. Enhancement of parents' awareness about β-Thalassemia disorder through two educational programs. Pak J Med Sci 2008; 24:283-6.
8. Paholpak S, Jetsrisuparb A, Wiangnon S, Sangsahachat D, Padtawaro LO. Result of video-education on "genetic transmission in thalassemia" thalassemic patients and their parents. J Med Assoc Thai 2006; 89:1909-14.
9. Ghanei M, Adibi P, Movahedi M, Khami MA, Ghasemi RL, Azarm T, et al. Pre-marriage prevention of thalassemia:report of a 100,000 case experience in Isfahan. Public Health 1997; 111:153- 6.
10. Tasleem S, Taseem H, Siddiqui MA, Adil MM, Rashid Y. Prenatal diagnosis of beta-thalassemia by chorionic villus sampling. J Pak Med Assoc 2007; 57:528-31.
11. Karimi M, Jamalian N, Yarmohammadi H, Askarnejad A, Afrasiabi A, Hashemi A. Premarital screening for β-thalassemia in Southern Iran: options for improving the programme. J Med Screen 2007; 14:62-66
12. Petrou M. Genetic Counselling. In: Prevention of Thalassaemias and Other Haemoglobin Disorders: Volume 1: Principles edited by J O, 2nd edition edn. Nicosia, Cyprus: Thalassaemia International Federation; 2013.
13. Cao A, Kan YW. The Prevention of Thalassemia. Cold Spring Harbor Perspectives in Medicine. 2013; 3(2).
14. Firdous N. Prevention of thalassaemia and haemoglobinopathies in remote and isolated communities--the Maldives experience. Ann Hum Biol. 2005;32(2):131–7.
15. Mitchell JJ, Capua A, Clow C, Scriver CR. Twenty-year outcome analysis of genetic screening programs for Tay-Sachs and beta-thalassemia disease carriers in high schools. Am J Hum Genet. 1996;59(4):793–8.
16. Dewanto JB, Tansah H, Dewi SP, Napitu H, Panigoro R, Sahiratmadja E. Increased Knowledge of Thalassemia Promotes Early Carrier Status Examination among Medical Students. Universa Medicina, 2016;34(3):220. doi.org/10.18051/UnivMed.2015.v34.220-228.
17. Miri-Moghaddam E, Motaharitabar EM, Erfannia E, Dashipour A, Oushvar M. High School Knowledge and Attitudes towards Thalassemia in Southeastern Iran. International Journal of Hematology-Oncology and Stem Cell Research, 2014;8(1):24-30. www.ncbi.nlm.nih.gov/pmc/articles/PMC3913153/#__ffn_sectitle.
18. Palash D, Srijana M, Kamalika D. Knowledge and Attitude Regarding Premarital Screening of Beta Thalassemia among The Students of Selected College of South-24-Parganas, West Bengal, India. India Journal Research, 2018;7(10):24-26. doi.org/10.36106/paripex.
19. Basu M. A Study on Knowledge, Attitude, and Practice about Thalassemia among General Population in Outpatient Department at A Tertiary Care Hospital of Kolkata. Journal of Preventive Medicine and Holistic Health, 2015;1(1):5-12. www.researchgate.net/publication/282657666
20. Kasper LD, Fauci AS, Longo DL, Braunwald E, Hauser SL, Jameson JL. Harrison's Principles of Internal Medicine (19th edition). 2015. United State: Mc Graw Hill. Pp 598-600.
21. Ebrahim S, Raza AZ, Hussain M, Khan A, Kumari L, Rasheed R, et al. Knowledge and Beliefs regarding Thalassemia in an Urban Population. Cureus, 2019; 11(7). doi.org/10.7759/cureus.5268.
22. Balcin YI, Ergin A, Polat A, Atilgan T, Uzun U, Koyuncu H. Thalassemia Premarital Screening Program: Public View. What has been Done, What Needs to be Done?. International Journal of Hematology and Oncology, 2014;24(4):247-252. doi.org/10.4999/uhod.14569
23. Hossain, M.S., Hasan, M.M., Raheem, E. et al. Lack of knowledge and misperceptions about thalassaemia among college students in Bangladesh: a cross-sectional baseline study. Orphanet J Rare Dis 15, 54 (2020). https://doi.org/10.1186/s13023-020-1323-y
24. Cheng K, Fucharoen S, Sanchaisuriya K, Fucharoen G, Sanchaisuriya P, Jetsrisuparb A. Effect of health education on severe thalassemia prevention and control in communities in Cambodia. Arch Public Health. 2018 Feb 19;76:13.
25. Muhammad L, Wajid KK, Afridi I, Ullah S, Khan A, Muhammad A. Awareness Among Educated and Uneducated Parents of Beta-Thalassemia Major Patients About Antenatal Screening. Cureus. 2022 Aug 29;14(8):e28561.
26. Usman, M., Moinuddin, M., Ghani, R., & Usman, S. (2009). Screening of five common beta thalassemia mutations in the Pakistani population: a basis for prenatal diagnosis. Sultan Qaboos University Medical Journal, 9(3): 305– 310.