Riset Informasi Kesehatan, Vol. 14, No. 1 (January 2025)
ISSN 2548-6462 (online), ISSN 2088-8740 (print)

DOI: 10.30644/rik.v14i1.964

Maternal knowledge to care for children with Beta-Thalassemia: Scoping Review

Hani Risnawati[1]*, Rantiningsih sumarni[2]
[1]fakultas ilmu kesehatan prodi keperawatan Universitas Duta Bangsa
[2]fakultas sains dan teknologi kesehatan prodi keperawatan Universitas Sahid Surakarta
*Corresponding author's email: hani_risnawati@udb.ac.id

Accepted: 11 January 2025; revision: 27 January 2025; published: 31 January 2025

Abstract

Introduction: Thalassemia, a genetic disorder prevalent in approximately 7% of the global population, poses significant health challenges, especially in Indonesia, where approximately 2,500 infants are diagnosed annually. The condition commonly manifests as beta-thalassemia, classified into major, intermedia, and minor forms, with varying clinical severity and treatment needs. Children with thalassemia often face chronic health problems that impact their quality of life, requiring lifelong care, including regular blood transfusions and iron chelation therapy. Parents, especially mothers, are crucial in managing thalassemia, as their knowledge and understanding of the disease significantly affect patient outcomes.
Purpose: to assess mothers' knowledge regarding the care of children with beta-thalassemia,
Methods: Search articles using four databases, namely Google Scholar, Wiley, Science Direct, and PubMed, using the PRISMA method using the keywords Maternal Knowledge to Care for Children with Beta-Thalassemia." 8 articles that met the inclusion criteria were selected from 9,950 articles in the initial.
Results: This review focuses on the correlation between mothers' educational background, understanding of thalassemia management, and the well-being of affected children. Findings suggest that inadequate maternal knowledge often results in suboptimal care practices, underscoring the need for improved educational programs and support structures to empower mothers to manage their children's health effectively. Therefore, improving maternal education regarding thalassemia may lead to better disease management practices, facilitate timely intervention, and ultimately improve the quality of life of children with this genetic disorder.
Conclusion: Maternal knowledge is crucial and necessary in both thalassemia disease and in-home care.

Keywords: Beta Thalassemia; Children with Beta Thalassemia; Maternal knowledge.

INTRODUCTION

Thalassemia is one of the most common genetic diseases in the world, with approximately 7% of the global population being carriers. In Indonesia, there are 10,531 thalassemia patients, and it is estimated that every year, around 2,500 babies are diagnosed with thalassemia. (1). Thalassemia can be divided into two types, namely alpha and beta thalassemia, of which beta thalassemia is the most commonly found type. Beta thalassemia is caused by the beta-globin chain's complete absence (B0) or partial reduction (B+). (2). Iron chelation therapy, regular red blood cell transfusions for transfusion-related iron overload, and supportive care to manage the consequences of iron overload are the hallmarks of treatment. In the absence of treatment, before the age of five, the majority of children with -thalassemia major are diagnosed1. This thalassemia is caused by total suppression of the beta-globin chain (B 0) or partial suppression of the beta-globin chain (b+). A compensatory increase in Hb A2 and Hb F occurs in all because the amount of excess alpha globin chain in homozygotes (Beta Thalassemia major) determines the severity of the following clinical manifestations, whereas heterozygotes (Beta Thalassemia minor) have little or no anemia and no symptoms.

Beta thalassemia is divided into three clinical forms: major, intermedia, and minor/trait carrier. Patients do not show symptoms and are only carriers in thalassemia minor, while thalassemia intermedia requires blood transfusions, although not as much as in thalassemia major. According to the Ministry of Health of the Republic of Indonesia (1), patients with thalassemia major must undergo regular blood transfusion therapy throughout their lives. Thalassemia is a disease that is inherited from parents to children. Children with thalassemia have carried the thalassemia gene since they were in the womb. In theory, clinical signs of thalassemia can be seen when the child is 1-2 years old. Children with thalassemia are generally more often found at school age (6-12 years) due to their worsening clinical condition, so most patients only get medical treatment when they are around 6 years old. (2).

Thalassemia beta is a lifelong genetic disease that requires long-term care. Social, psychological, and physical conditions can affect the chronic state of the child, which in turn impacts their quality of life (3). The clinical manifestations of thalassemia are anemia, weight loss, pallor, weakness, and flat nose. Suppose these clinical manifestations of thalassemia are not considered. Then, the condition of thalassemia patients will impact their quality of life. Complications in thalassemia patients are heart, bone, liver, and hormone gland complications. Management in thalassemia is non-pharmacological and pharmacological. Non-pharmacology consists of food with balanced nutrition, pharmacological transfusion, and administration of iron chelation therapy drugs (4). Complications in thalassemia can occur due to the disease itself, side effects of treatment, or iron chelation therapy. Some complications that may arise include heart failure, growth disorders, enlarged spleen and liver, delayed puberty, and others. The success of treatment and prevention of complications in children with thalassemia is highly dependent on the support provided by parents (3).

Parents are responsible for children and play an essential role in treating children with thalassemia. This is due to the need for lifelong medication and treatment to prevent complications that can affect the quality of life of children with thalassemia. Therefore, parental support in treating children with thalassemia is needed to improve their quality of life. (4). This is in line with research conducted by (5) The results of her research show that family relationships are indispensable in improving the quality of life of children with thalassemia in the children's room.

Good knowledge of how to treat and manage this condition is essential. Maternal knowledge includes an understanding of the disease itself, medication management, and strategies to prevent complications that may arise. Many studies have shown that there is still a lack of adequate understanding among mothers, whether regarding the medical aspects of the disease, nutritional needs, or the psychosocial impact that the child and family may experience. Factors such as limited access to information, education, and social support affect the extent to which mothers can acquire and apply appropriate knowledge in caring for children with beta-thalassemia(6).

METHOD

A. Search Design and Methods

This literature review search came from three databases: Google Scholar, Wiley, Science Direct, and PubMed Journals published between 2019 and 2024. The process of extracting this review article used specific clinical questions, namely with PCC [Population (P), Concept (C) and Context (C)]. Population (P) is children with thalassemia, Concept (C) is related to Maternal knowledge to care for children with beta-thalassemia, and Context (C) is community setting. We used several keywords or keywords with a Boolean search, namely Maternal Knowledge to Care for

Children with Beta Thalassemia. The selection of articles in this study follows a method called Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA).

B. Identifying Research Question

This literature review study aims to answer the following questions: how is the mother's knowledge related to care in children with beta-thalassemia to determine the feasibility of the question based on the review for this study used population, exposure, and outcome?

C. Inclusion and Exclusion Criteria

The inclusion criteria in this study were articles: (1) articles that explain feeding rules; (2) research articles within the last 5 years: 2018-2023; (3) open access articles, full text; (4) Original articles or research articles. Exclusion criteria in this study included articles that were protocol studies. All authors performed the first screening and content analysis of the articles.

D. Quality Appraisal

Before conducting the quality appraisal of the articles, the writing team read the ten selected full-text articles. After the team finished reading the articles, the team conducted a critical appraisal using the Joanna Briggs Institute (JBI) critical appraisal checklist for analytical cross-sectional, case-control, randomized controlled trials, quasi-experimental, cohort, and qualitative studies. Ten articles were assessed for quality using the JBI critical appraisal checklist, and all articles met the criteria. No articles were excluded based on quality appraisal

RESULTS

A. Search Results

The team's search results were found in the Google Scholar database, Science Direct Pubmed: 9950. Screening based on the year and duplicate articles left 5192 articles. Then, the screening process of titles, abstracts, and full-text availability left 59 articles, and screening based on inclusion and exclusion criteria (eligibility criteria) left 13 articles. Furthermore, the article carried out a quality assessment through the JBI critical appraisal, and five articles were excluded so that the remaining eight articles could be analyzed (Figure 1).

B. Data Extraction and Analysis

Eight articles were extracted and analyzed in this stage to identify authors, year of publication, country of origin, purpose, population and sample size, methods, and significant findings based on review questions.

Picture 1. PRISMA Flowchart

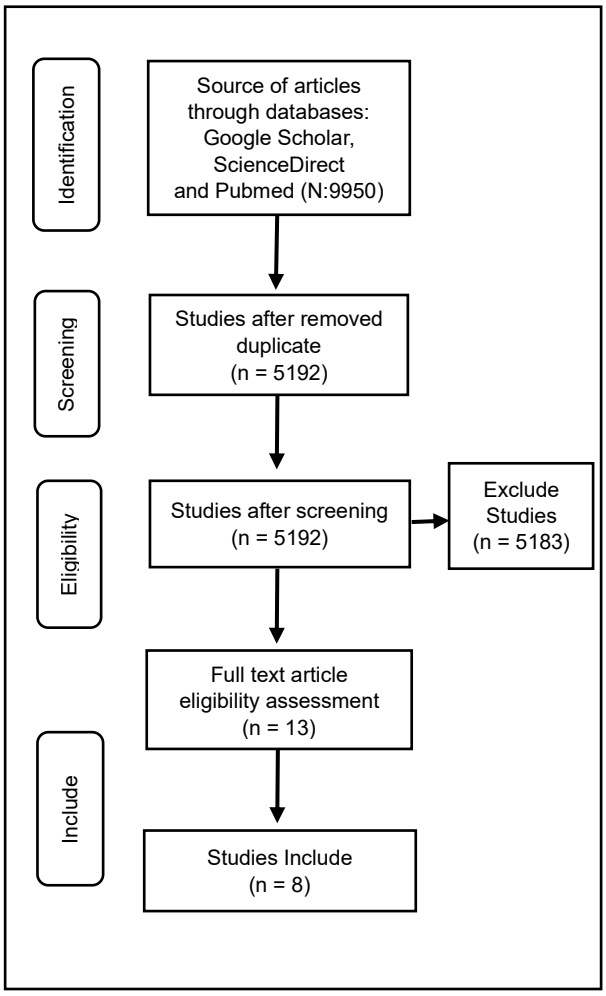

Table 1. General information of the seven included articles

No	Author	Year	Country	Continent
1.	Nandeswari *et al.*	2021	Indonesia	Asia
2.	Riskawaty *et al*	2023	Indonesia	Asia
3.	Faraski *et al*	2023	Indonesia	Asia
4.	Andriani *et al*	2022	Indonesia	Asia
5.	Karakul *et al*	2022	Turkey	Asia
6.	Atshan and Aziz	2022	Iraq	Asia
7.	Mahfoz *et al.*	2024	Egypt	Asia
8.	Brair and Mohammad	2020	Jordan	Asia

Table 2. Articles synthesis

No	Author, Year	Research Objective	Research Design	Setting
1	Nandeswari *et al,* 2021	Knowing the description of the level of knowledge of parents who care for children with thalassemia.	Quantitative non-experimental research design with descriptive survey design.	Parents of the POPTI Association of Schools aged 6-12 years. 51 parents with thalassemia children at the Palembang branch of the Indonesian Thalassemia Parents Association
2	Riskawaty *et al*, 2023	Knowing family knowledge about thalassemia child care at home	Quantitative research with crossectional	Population with total sampling of children with thalassemia aged 1-20 years Pediatric clinic of NTB Provincial Hospital
3	Faraski *et al.*, 2023	Knowing the relationship between education level and parental knowledge with quality of life in thalassemia children.	Quantitative research with cross-sectional	52 respondents of parents of children with thalassemia at RSUD Dr. M. Yunus Bengkulu
4	Andriani *et al*, 2022	to deeply understand the picture of fortitude that focuses on the mother's experience in caring for a child with thalassemia. Maternal care efforts, commitment, control and challenges in caring for children with thalassemia.	Qualitative research with phenomenological experience	6 parents with thalassemia children in Indonesia
5	Karakul *et al*, 2022	To identify issues related to the disease process of 12-18-year-olds with beta-thalassemia major and to test the effectiveness of individualized empowerment education about these needs.	Mixed-type study	30 children with beta-thalassemia aged 12-18 years in a thalassemia unit hospital in Turkey
6	Atshan and Aziz, 2022	To determine the effect of the education program on parents' knowledge about chelation therapy and nutrition for their children with thalassemia beta major.	A quasi-experiment study	35 families with children with thalassemia major in Al Najif region
7	Mahfoz *et al.,* 2024	Knowing the effects of a stepped program phased program on mothers' care of their children with beta-thalassemia.	Pre-experimental design	115 mothers with thalassemia children and no mental retardation, congenital diseases, and other chronic diseases
8.	Brair and Mohammad, 2020	Knowing the knowledge and care practices of mothers of pediatric thalassemia children in Jordan.	Cross-sectional design	General hospitals in 3 regions of Jordan

Table 3. Maternal Knowledge

No	Author, Year	Research Outcomes
1.	Nandeswari *et al*., 2021	The results showed that most of the knowledge levels of parents who care for children with thalassemia are categorized as less, namely as many as eight people, with a percentage of 47.1%.
2.	Riskawaty *et al*, 2023	Spearman rank test results with a correlation value of .453 with a p-value of 0.004, meaning <0.05 Ha is accepted; there is a relationship between the family knowledge level of home care in Thalasemia patients.
3.	Faraski *et al.*, 2023	There is a significant correlation between parents' level of education and knowledge and the quality of life of children with thalassemia major.
4.	Andriani *et al*, 2022	The themes identified from this study were not accepting reality, resignation, attempts to treat the child through medical and traditional means, challenges, and maternal fortitude. We can conclude that a mother's fortitude in caring for a child with thalassemia can be successful if the mother accepts reality. This can be seen and felt through the mother's patience, sincerity, and prayer in caring for her child.
5.	Karakul *et al*, 2022	Individualized education and empowerment positively affect the child's knowledge of the disease, treatment process, mother-father relationship, and social life needs.
6.	Atshan and Aziz, 2022	The educational program was highly effective in enhancing parents' knowledge in the study group, with an increase in the mean score of parents' knowledge through post-tests one and 2. There was a statistically significant difference—the relationship between parental knowledge and affected virus-carrying fathers and mothers.
7.	Mahfoz *et al.,* 2024	The program improved maternal knowledge, practices, and attitudes regarding beta-thalassemia. It can be recommended that the program for mothers of children with thalassemia be implemented in all health facilities serving this patient population.
8.	Brair and Mohammad, 2020	Most mothers had a low level of knowledge regarding thalassemia, and most incorrectly practiced Desferal tablet administration. No significant association was found between mothers' knowledge and care practices of mothers and their socio-demographic characteristics.

DISCUSSION

Maternal knowledge in children with thalassemia is proportional to the level of education. So, the more parents understand about caring for children with thalassemia, the better the quality of life of children with thalassemia also increases. Government programs and health workers are critical in providing information, teaching how to handle it at home, and knowledge about thalassemia disease. Parents' knowledge is essential in facilitating the decision-making and actions children with thalassemia need. The success of treating and caring for children with thalassemia is related to parents' knowledge.

Thalassemia is an inherited hemoglobin condition characterized by decreased or missing globin formation, unbalanced buildup of globin chains, and inefficient erythropoiesis with hemolysis. Based on the results of research by (14) , there is a significant relationship between the problems that arise in thalassemia patients and parental knowledge. Parents' knowledge in caring for thalassemia patients at a certain age can facilitate decision-making and actions needed if the patient requires care at home or in the hospital. (14). Parents need good

knowledge about thalassemia disease, the treatment required, scheduling blood transfusion therapy, and managing the care of thalassemia patients at home to reduce symptoms. Research by (15) It also states that parents with good knowledge are expected to carry out their duties to care for thalassemia patients, improving their quality of life.

The results of (16) This states that suboptimal knowledge in parents of children with thalassemia includes thalassemia, which is a chronic disease that is inherited, and thalassemia children can experience growth and development disorders, including delayed puberty. Respondents also made many mistakes in the indicators of questions regarding activity and rest in thalassemia children. According to (15), parents' lack of knowledge about thalassemia disease can result in suboptimal care provided by parents to children with thalassemia. Research by (17) States that the level of parental knowledge about thalassemia disease is related to the non-optimal treatment undertaken by children, including the timeliness of transfusion and consumption of iron chelation drugs.

These results agree with a study done in Egypt, which researched to determine the impact of an educational program about iron chelation therapy in Egypt. It demonstrates that management had the lowest percentage of satisfactory knowledge among the studied parents before the program. Statistically significant improvements were revealed in the post-intervention phase in all knowledge domains. There was a slight decrease in the follow-up, especially in management (54.0 %). However, the percentages of satisfactory knowledge remained significantly higher compared to pretest. The findings of Karen’s knowledge about their age agree with the study, which aimed to increase parents' knowledge and practice by establishing a health education program in Egyptian.

The present study's results found inadequate knowledge (60%) and poor care practice (71.1%) among mothers of thalassemic children in Jordan. These findings are consistent with other studies from developing countries that reported inadequate knowledge regarding beta-thalassemia and its management among parents and the general public, which could negatively affect the health and safety of children with thalassemia. (18),(19). This could be explained by the absence of structured educational programs for thalassemic patients and their guardians in Jordan. Basu, in 2015, found only 14.02% had good practices towards thalassemia among participants and, as a result, recommended awareness programs about the management of thalassemia to be offered to the general public. (19).

CONCLUSION

The level of maternal knowledge about thalassemia dramatically influences how a child with thalassemia is managed and treated. Good knowledge can help in early detection, disease management, preventing complications, and providing the child with a better quality of life. Therefore, it is essential to improve education and information about thalassemia to mothers, either through public health programs, genetic counseling, or easier access to medical information.

REFERENCES

1. Kementrian Kesehatan Republik Indonesia. (2019). Angka Pembawa Sifat Talasemia Tergolong Tinggi. Retrieved 7 Januari 2025, from Kementrian Kesehatan Republik Indonesia: www.p2ptm.kemenkes.go.id
2. Xie J, Zhou Y, Xiao Q, Long R, Lianxiang L, Lei L. Rare double heterozygosity for poly a(A) g) and cd17(a> t) of beta thalassemia intermedia in a chinese family.

Hematol Rep. 2019;11(3):52–4.

3. Nandeswari V, Indriansari A, Latifin K. Pengetahuan Orang Tua Merawat Anak Penderita Thalasemia. J Keperawatan Fak Kedokt Univ Sriwij. 2021;155–62.
4. Cappellini, MD et al. Guidelines for theManagementofTransfusi darahon Dependent Thalassemia (TDT), 3rd ed. Nicosia: ThalassaemiaInternasional Federation;2016
5. Pranajaya, R., & Nurchairina. (2016). Faktor yang Berhubungan dengan Kualitas Hidup Anak Thalasemia. Jurnal Keperawatan, XII(1), 130-139.
6. Sawitri H, Husna CA. Karakteristik Pasien Thalasemia Mayor Di Blud Rsu Cut Meutia Aceh Utara Tahun 2018. AVERROUS J Kedokt dan Kesehat Malikussaleh. 2018;4(2):62.
7. Riskawaty, H. M., Wasliah, I., & Ramadani, N. I. (2023). Hubungan tingkat Pengetahuan Keluarga Terhadap Perawatan Di Rumah Pada Penderita Thalasemia RSUD Provinsi NTB. 3, 9644–9655.
8. Faraski, K., Rohima, W., & Putri, S. R. (2023). Hubungan Antara Tingkat Pendidikan dan Pengetahuan Orangtua dengan Kualitas Hidup Anak Talasemia Mayor di RSUD Dr. M. Yunus Bengkulu. Syntax Idea, 5(11), 1992-2011. https://doi.org/10.46799/syntax-idea.v5i11.2705
9. Andriani, R., Nurhaeni, N., & Safariyah, E. (2022). Mother' s Fortitude in Taking Care of Children with Thalassemia in Indonesia. 2022, 574–584. https://doi.org/10.18502/kls.v7i2.10358
10. Karakul, A., & Şenol, S. (2022). Effectiveness Of Individualized Empowerment Education On Children With Beta-Thalassemia Major: A Mixed-Method Study. Izmir Democracy University Health Sciences Journal, 5(3), 720-734.
11. Atshan, R. S., & Radha, A. (2022). Effectiveness of an Educational Program on Parents ' Knowledge about Home Health Care Management to Children with Beta Thalassemia-Major at Thalassemia Center in Al-Zahra Teaching Hospital for Maternity and Children in Al-Najaf City. April. https://doi.org/10.53350/pjmhs22163931.
12. Fathi Ahmed Mahfoz, F., Hassan Mahmoud, E., & Abdelhamid Zaki, N. (2024). Effect of Stepwise Program on Mothers' Care of their Children with Beta Thalassemia. Egyptian Journal of Health Care, 15(3), 449-461.
13. Khresheh, S. M., Brair, S. L., Knowledge, K., & Practices, C. (2020). Knowledge and practices among Mothers about Care of their children with Beta Thalassemia Major : A descriptive Study. 11(3).
14. Marnis, D., Indriati, G., & Nauli, F. (2018). Hubungan Tingkat Pengetahuan Ibu dengan Kualitas Hidup Anak Thalasemia. Jurnal Keperawatan Sriwijaya, 5(2), 31-41.
15. Ishfaq, K., Ahmad, T., Naeem, S., Ali, J., & Zainab, S. (2016). The Knowledge of Parents Having Thalassemia Child. Isra Medical Journal, 8(2), 79-82.
16. Supriyanti, E., & Mariana, M. (2019). Faktor-Faktor yang Berhubungan dengan Kepatuhan Transfusi pada Pasien Thalasemia. Jurnal Ilmiah Ilmu Keperawatan Indonesia, 9(2), 607-615.
17. Indanah, I., Yetti, K., & Sabri, L. (2012). Pengetahuan dan Dukungan Sosial Meningkatkan Self-Care Behavior pada Anak Sekolah dengan Thalasemia Mayor. Jurnal Keperawatan Indonesia, 15(1), 53-60.

18. Saxena, A. (2015). Growth Retardation in Thalassemia Major Patient . Int J Hum Genet.
19. Biswas, B., Naskar, N., Basu, R., Dasgupta, A., Paul, B., & Basu, K (2018). Knowledge the Caregivers of Thalassemic Children Regarding Thalassemia: A Cross-sectional Study in a Tertiary Care Health Facility of Eastern India. Iraqi Journal of Hematology , 49-54