



## Caring for Thalassemia Patients: The Nursing Role and Its Impact

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### Abstract:

Caring for thalassemia patients requires a multifaceted approach, as the condition primarily affects hemoglobin production and leads to a myriad of complications. Nurses play a crucial role in managing these patients throughout their treatment journey. They assess and monitor the clinical status of patients, administer blood transfusions, and ensure that iron chelation therapy is properly implemented to prevent iron overload, a common consequence of repeated transfusions. Additionally, nurses provide essential patient education regarding lifestyle modifications, dietary considerations, and the importance of regular follow-up appointments, thereby empowering patients and their families to better manage the disease. Their ability to recognize emotional distress and facilitate psychosocial support also contributes significantly to the overall well-being of these patients. The impact of nursing care in the management of thalassemia can be profound, as effective nursing interventions contribute to improved patient outcomes and quality of life. Through collaboration with interdisciplinary teams, nurses advocate for the needs of thalassemia patients, ensuring that they receive comprehensive care tailored to their specific circumstances. The therapeutic relationship that nurses build with patients fosters trust and encourages adherence to treatment regimens. Moreover, by engaging in community outreach and awareness programs, nurses can educate the public on thalassemia, promote early screening, and encourage genetic counseling, ultimately playing a vital role in reducing the prevalence of the disorder. In essence, the dedication and expertise of nursing professionals are pivotal in transforming the care experience for those living with thalassemia.

## 1. Introduction

Thalassemia, a group of inherited hemoglobinopathies characterized by defective synthesis of globin chains, represents a significant global health challenge with a profound personal and societal impact. As one of the most common monogenic disorders worldwide, it poses a lifelong burden on those diagnosed, requiring complex and continuous medical management to ensure survival and quality of life. The World Health Organization (WHO) estimates that approximately 1.5% of the global population are carriers of beta-thalassemia, with around 60,000 symptomatic individuals born annually, primarily in the Mediterranean, Middle East, Transcaucasus, Central and South Asia, and North Africa regions [1].

The advent of regular blood transfusion therapy revolutionized the prognosis for thalassemia major patients, transforming it from a fatal pediatric disease into a chronic condition with a life expectancy that now extends well into adulthood. However, this life-sustaining therapy comes with a formidable consequence: chronic iron overload. Each unit of transfused blood contains approximately 200-250 mg of iron, which the human body has no active mechanism to excrete. This cumulative iron deposition in vital organs such as the heart, liver, and endocrine glands becomes the primary cause of morbidity and mortality in this population. The introduction of iron chelation therapy in the 1970s marked a second revolutionary step, directly countering this iatrogenic complication and further extending life expectancy [2]. Despite these medical advancements, the journey for a thalassemia patient remains a relentless cycle of clinic visits, transfusions, chelation, and monitoring for complications, making it a paradigm of a complex chronic illness.

Within this challenging landscape, the role of nursing has evolved from a task-oriented, supportive function to a central, multifaceted, and dynamic pillar of comprehensive care. The contemporary nurse in thalassemia management is no longer merely an executor of physician orders but is a core member of the multidisciplinary team, acting as a clinician, educator, coordinator, counselor, and patient advocate. The complexity of modern thalassemia care, which encompasses the management of advanced drug regimens, sophisticated infusion pumps, and a wide array of potential complications from endocrinopathies to cardiac dysfunction, demands a highly specialized and knowledgeable nursing workforce [3].

The nursing impact begins at the point of diagnosis, often providing crucial support and education to

distressed families, and extends across the entire lifespan of the patient. Nurses are the frontline professionals responsible for the safe administration of blood products, vigilantly monitoring for and managing acute transfusion reactions, which range from mild febrile responses to life-threatening anaphylaxis. They are the primary educators teaching patients and their families the techniques and importance of adherence to iron chelation therapy, whether it be subcutaneous infusions of deferoxamine or the complexities of oral agents like deferasirox and deferiprone, including their side-effect profiles and drug interactions [4]. Studies have consistently shown that adherence to chelation therapy is the single most critical modifiable factor in preventing iron-induced organ damage and premature death, placing the nurse's educational and motivational role at the heart of long-term patient outcomes [5].

Furthermore, the nursing role is pivotal in the surveillance and early detection of complications. Nurses perform essential assessments, track ferritin and other biomarker trends, and are often the first to identify subtle signs of cardiac siderosis, endocrine dysfunction like hypothyroidism or diabetes, or hepatic fibrosis. They serve as the crucial link between the patient and other specialists, ensuring a coordinated and seamless care experience. Beyond the physical domain, the psychological and social burden of thalassemia is immense. Patients frequently grapple with issues of body image related to disease manifestations or chelation pump sites, anxiety about the future, depression, and challenges in social integration, education, and employment [6]. The nurse, through therapeutic communication and sustained patient relationships, provides invaluable psychosocial support, identifies mental health needs, and facilitates referrals to appropriate services.

The evidence base supporting the transformative impact of specialized nursing care on thalassemia outcomes is growing. Research indicates that structured nurse-led education programs significantly improve patient knowledge, self-management skills, and adherence to chelation therapy, directly correlating with lower serum ferritin levels—a key surrogate marker for iron burden [7, 8]. Nurse-led clinics and transition programs for adolescents moving from pediatric to adult care have been shown to improve clinic attendance, patient satisfaction, and health-related quality of life (HRQoL) scores [9]. The holistic, patient-centered approach championed by nursing is essential for addressing the multifaceted challenges of this chronic condition.

### **Thalassemia as a Chronic Condition**

Thalassemia represents one of the most common monogenic disorders worldwide, posing a significant public health challenge in many regions. According to recent epidemiological data from the Thalassaemia International Federation, approximately 1.7% of the global population are carriers of beta-thalassemia, with about 60,000-70,000 symptomatic children born annually worldwide [13]. The disease demonstrates a distinctive geographical distribution pattern, with high prevalence in the Mediterranean basin, Middle East, Indian subcontinent, Southeast Asia, and Africa, though globalization and population mobility have made it a concern in virtually every country. The inheritance follows an autosomal recessive pattern, meaning both parents must be carriers for a child to be affected, with each pregnancy having a 25% chance of producing an affected child, a 50% chance of producing a carrier, and a 25% chance of producing an unaffected child [14].

The fundamental pathophysiology of thalassemia involves genetic mutations that disrupt the synthesis of globin chains, leading to imbalanced production of alpha and beta chains. In beta-thalassemia, which represents the most severe form, there is reduced or absent production of beta-globin chains. This imbalance results in unpaired alpha-globin chains that precipitate in erythroid precursors in the bone marrow and peripheral blood, causing ineffective erythropoiesis and hemolytic anemia. The clinical spectrum ranges from the severe transfusion-dependent thalassemia major to the milder non-transfusion-dependent thalassemia intermedia, with various intermediate forms [15]. The severity of clinical manifestations directly correlates with the degree of globin chain imbalance, which is determined by the specific genetic mutations inherited.

The introduction of regular blood transfusion therapy in the mid-20th century transformed thalassemia major from a universally fatal disease in childhood to a chronic manageable condition. Before this therapeutic advancement, children with thalassemia major typically succumbed to severe anemia or cardiac complications within the first few years of life. The current standard of care involves maintaining a pre-transfusion hemoglobin level above 9-10.5 g/dL, which suppresses ineffective erythropoiesis, allows for normal growth and development in children, and enables patients to maintain reasonable energy levels and quality of life [16]. Regular transfusions, typically administered every 2-4 weeks, have become the lifeline that sustains patients with thalassemia major throughout their lives.

However, this life-sustaining therapy carries a formidable burden in the form of chronic iron overload. The human body lacks an active mechanism to excrete excess iron, and each unit of transfused blood contains approximately 200-250 mg of iron. Consequently, patients receiving regular transfusions accumulate 0.4-0.5 mg of iron per kg of body weight per day, leading to total body iron stores that can reach 40-50 grams by early adulthood – more than ten times the normal iron content [17]. This iron deposition follows a characteristic pattern, primarily affecting the liver, heart, and endocrine glands, where it generates toxic hydroxyl radicals through the Fenton reaction, causing oxidative damage to cellular structures including proteins, lipids, and DNA.

The clinical consequences of iron overload are devastating and represent the primary cause of morbidity and mortality in thalassemia patients in the modern era. Cardiac iron overload leads to cardiomyopathy, arrhythmias, and ultimately heart failure, which remains the leading cause of death despite therapeutic advances. Hepatic iron deposition causes fibrosis, cirrhosis, and potentially hepatocellular carcinoma. Endocrine complications are equally widespread and debilitating, including hypogonadism leading to delayed or absent puberty, growth retardation, diabetes mellitus, hypothyroidism, and hypoparathyroidism [18]. The cumulative impact of these complications creates a complex clinical picture that requires multidisciplinary management throughout the patient's life.

The introduction of iron chelation therapy in the 1970s marked the second revolutionary advancement in thalassemia care, directly addressing the iatrogenic complication of transfusion-induced iron overload. The first available chelator, deferoxamine, required subcutaneous infusion over 8-12 hours, 5-7 days per week, presenting significant challenges to patient adherence and quality of life. The subsequent development of oral chelators, including deferiprone and deferasirox, has expanded therapeutic options and improved convenience, though each agent carries its own profile of efficacy and potential adverse effects [19]. The timing of chelation initiation is critical, typically beginning after 10-20 transfusions or when serum ferritin levels consistently exceed 1000 ng/mL, with the goal of maintaining serum ferritin below 1000-1500 ng/mL to prevent organ damage [20].

### **Role of the Modern Thalassemia Nurse**

The foundation of the modern thalassemia nurse's practice is built upon specialized clinical expertise.

This goes beyond the technical skill of venipuncture or intravenous cannulation for transfusion therapy. It involves a deep understanding of the disease's pathophysiology, the pharmacology of iron chelation agents, and the nuanced presentation of disease complications. Nurses are responsible for conducting comprehensive patient assessments before each transfusion, evaluating not only vital signs but also subtle indicators of disease progression or treatment complications, such as signs of heart failure, jaundice, or endocrine dysfunction [21]. During transfusions, they exercise vigilant monitoring for both acute and delayed reactions, possessing the clinical judgment to distinguish between a mild febrile non-hemolytic reaction and a potentially life-threatening anaphylactic or hemolytic event. Their expertise ensures that the life-sustaining therapy of transfusion is delivered with maximum safety and efficacy.

Perhaps one of the most critical roles of the thalassemia nurse is that of an educator and adherence promoter. The success of thalassemia management hinges critically on long-term adherence to complex treatment regimens, particularly iron chelation therapy. Nurses are the primary professionals responsible for educating patients and their families about the rationale for chelation, the consequences of non-adherence, and the practicalities of administration. This education is not a one-time event but an ongoing, adaptive process. For patients on subcutaneous deferoxamine, nurses teach and assess proper infusion pump use, rotation of injection sites to prevent lipodystrophy, and management of local reactions. For those on oral agents like deferasirox or deferiprone, they provide detailed instruction on timing, dietary considerations, and monitoring for specific side effects such as gastrointestinal disturbances, arthropathy, or neutropenia [22]. Studies have consistently demonstrated that structured, nurse-led education programs significantly improve medication adherence rates, which directly correlates with better-controlled serum ferritin levels and reduced organ iron concentration [23].

Functioning as a care coordinator and navigator represents another indispensable dimension of the nursing role. Thalassemia patients typically interact with numerous specialists throughout their lives, including hematologists, cardiologists, endocrinologists, hepatologists, and psychologists. The nurse acts as the central hub of communication, ensuring that information flows seamlessly between these different providers and that the care plan is cohesive and not fragmented. They coordinate appointments, facilitate referrals, and ensure that

relevant investigation results are available to all members of the multidisciplinary team. This coordination is especially vital during transitional periods, such as when an adolescent patient moves from pediatric to adult care services. Nurse-led transition programs have been shown to significantly improve continuity of care, reduce loss to follow-up, and empower young adults to become more autonomous in managing their health [24].

The psychosocial dimension of the nursing role cannot be overstated. Thalassemia imposes a heavy psychological burden on patients and their families. From an early age, patients grapple with the reality of a lifelong, life-limiting condition. They may experience anxiety about their future, depression, frustration with the demanding treatment schedule, and body image issues related to disease manifestations (e.g., bony deformities, growth retardation) or treatment devices (e.g., infusion pumps). The nurse, by virtue of their sustained and trusting relationship with the patient, is uniquely positioned to provide emotional support, screen for psychological distress, and employ therapeutic communication techniques to help patients and families develop effective coping strategies [25]. They create a safe environment where patients can express their fears and frustrations, which is fundamental to maintaining mental well-being and, by extension, treatment engagement.

Finally, the role of the nurse as a patient advocate is paramount. Thalassemia patients may face challenges ranging from insurance coverage for expensive medications to discrimination in educational or occupational settings. Nurses often serve as their advocates, liaising with insurance companies, providing necessary documentation for schools or employers, and empowering patients to assert their rights and needs within the healthcare system and broader society [26]. They ensure that the patient's voice is heard and that care is aligned with the patient's individual values, preferences, and life goals.

In conclusion, the role of the nurse in thalassemia care has expanded into a complex, integrated portfolio of responsibilities that are essential for addressing the multifaceted challenges of this chronic condition. By synthesizing clinical expertise with skills in education, coordination, counseling, and advocacy, the modern thalassemia nurse moves beyond task-oriented care to deliver a holistic, patient-centered approach. This evolved role is not merely supportive but is fundamentally instrumental in translating medical advances into tangible improvements in patient survival, complication prevention, and overall quality of life, establishing the nurse as an indispensable pillar of

the thalassemia multidisciplinary team [27, 28, 29, 30].

### The Nurse's Critical Role in Transfusion Therapy and Complication Management

The transfusion process begins long before the blood unit enters the patient's vein, with a meticulous pre-transfusion nursing assessment. This assessment is a holistic evaluation designed to ensure the patient is physiologically and psychologically prepared to receive the blood product. The nurse systematically reviews the patient's most recent laboratory results, paying particular attention to hemoglobin and hematocrit levels to confirm the clinical indication for the transfusion, as well as renal and hepatic function, which can influence transfusion tolerance. A thorough physical examination is conducted, assessing for signs of fluid overload, occult infection, or any new symptoms that might contraindicate transfusion or necessitate pre-medication. Crucially, the nurse verifies the patient's identity and cross-matches the blood unit against the prescription and patient identification with a rigorous, double-check protocol—a fundamental safety step to prevent the catastrophic consequence of an ABO-incompatible transfusion [31]. Furthermore, this pre-transfusion interaction provides a vital opportunity to assess the patient's and family's understanding of the procedure, address any anxieties, and reinforce education about potential reactions and the importance of reporting symptoms promptly.

Once the transfusion is initiated, the nurse enters a phase of intense, proactive monitoring. The first 15 minutes of any transfusion are the most critical, as the majority of severe acute reactions occur during this window. The nurse remains with the patient, closely observing for any signs of a reaction, which can range from subtle urticaria or flushing to more ominous symptoms like rigors, fever, respiratory distress, hypotension, or lumbar pain—a classic symptom of hemolysis. Standard monitoring protocols dictate frequent vital sign assessments (e.g., every 15 minutes for the first hour) and constant visual surveillance [32]. Beyond monitoring for reactions, the nurse manages the flow rate, adjusting it to ensure the blood is infused within the safe, prescribed timeframe (typically 2-4 hours per unit) while considering the patient's cardiac status and risk for fluid overload. This balancing act requires sophisticated clinical judgment, especially in vulnerable populations like young children or patients with pre-existing cardiac siderosis.

The ability to rapidly recognize and manage transfusion reactions is a cornerstone of the

thalassemia nurse's clinical competence. Reactions are broadly categorized into acute (occurring within 24 hours) and delayed (occurring days to weeks later). The most common acute reactions include febrile non-hemolytic transfusion reactions (FNHTR) and mild allergic reactions, which the nurse typically manages by temporarily stopping the transfusion, administering antipyretics or antihistamines as per protocol, and, once symptoms resolve, cautiously restarting the transfusion at a slower rate. However, the nurse must be perpetually vigilant for the signs of more severe, life-threatening reactions. These include:

- **Acute Hemolytic Reaction:** Often due to ABO incompatibility, presenting with fever, chills, back pain, hypotension, and hemoglobinuria. The nurse's immediate action is to stop the transfusion, maintain intravenous access with normal saline, and initiate emergency protocols [33].
- **Transfusion-Related Acute Lung Injury (TRALI):** A non-cardiogenic pulmonary edema presenting with severe dyspnea and hypoxia during or within 6 hours of transfusion. Management focuses on respiratory support.
- **Anaphylactic Reaction:** Characterized by bronchospasm, laryngeal edema, and hypotension, requiring immediate administration of epinephrine and advanced life support.

For delayed reactions, such as Transfusion-Associated Graft-versus-Host Disease (TA-GVHD) or post-transfusion purpura, the nurse's role shifts to one of surveillance and patient education, teaching patients to recognize and report late-onset symptoms like rash, diarrhea, or bruising [34].

A particularly insidious long-term complication of chronic transfusion therapy, aside from iron overload, is alloimmunization—the development of antibodies against foreign red blood cell antigens. This can make finding compatible blood increasingly difficult and heightens the risk of hemolytic reactions. Nurses play a pivotal role in alloimmunization prevention by meticulously documenting any transfusion reactions, ensuring that this information is permanently recorded in the patient's record, and advocating for the use of phenotypically matched blood units, especially for patients from ethnic minorities who may have antigen profiles that differ from the predominantly Caucasian blood donor pool [35].

Finally, the nurse's role in transfusion therapy is integral to patient education and empowerment. They educate patients and families about the signs and symptoms of both acute and delayed transfusion reactions, empowering them to be

active participants in their own safety. They explain the importance of infectious disease screening for blood products while contextualizing the minimal residual risk, helping to alleviate anxiety. For patients transitioning to self-management, nurses teach the skills of symptom recognition and reporting, fostering independence and self-efficacy [36].

In conclusion, the nurse's role in transfusion therapy for thalassemia is a dynamic and highly skilled practice that blends technical proficiency with sharp clinical judgment, vigilant monitoring, and effective patient communication. From the initial assessment to the management of complex reactions and long-term surveillance, the nurse acts as the patient's guardian, ensuring that their essential lifeline of blood is delivered with the highest possible degree of safety. This critical function not only prevents immediate harm but also safeguards the long-term viability of transfusion as a therapeutic strategy, directly contributing to the patient's longevity and quality of life [37, 38, 39, 40].

### **The Adherence Champions: Nursing Strategies for Effective Iron Chelation Therapy Management**

If regular blood transfusions represent the lifeline for patients with thalassemia major, then consistent and effective iron chelation therapy constitutes the essential safeguard that protects them from the toxic consequences of that lifeline. The management of iron overload through chelation is arguably the most complex, demanding, and psychologically taxing aspect of long-term thalassemia care. It is within this challenging domain that the role of the thalassemia nurse evolves from clinician to "adherence champion"—a dedicated professional who employs a multifaceted arsenal of strategies to educate, motivate, support, and problem-solve with patients to overcome the significant barriers to long-term adherence. The success of this nursing role is directly measurable in the surrogate markers of iron burden and, ultimately, in the prevention of end-organ damage and the extension of life.

The foundation of effective chelation management is a deep and nuanced understanding of the available pharmacologic options, each with its distinct profile of administration, efficacy, and side effects. Nurses must be experts in the three primary iron chelators: the subcutaneous infusion of deferoxamine (DFO), and the oral agents deferiprone (DFP) and deferasirox (DFX). For patients on DFO, the nursing role is intensely practical and educational. They are responsible for teaching patients and families the intricate process

of reconstituting the drug, operating the portable infusion pump, and performing subcutaneous needle insertion with proper site rotation to prevent skin complications like lipodystrophy or localized reactions. This requires hands-on training, return demonstrations, and ongoing assessment of technique [41]. For the oral agents, the focus shifts to managing the logistics of daily life. With deferasirox, nurses emphasize the importance of taking the medication on an empty stomach, the specific instructions for dispersing the tablet in water or juice, and the avoidance of concurrent aluminum-containing antacids. For deferiprone, they educate about the strict three-times-daily dosing schedule and the critical need for weekly neutrophil monitoring to detect agranulocytosis [42]. This detailed, drug-specific knowledge allows the nurse to provide tailored, accurate education that is fundamental to safe and effective use.

However, knowledge alone is insufficient to ensure adherence. The thalassemia nurse must adeptly identify and address the multifaceted barriers that patients face. These barriers are seldom purely clinical; they are often woven into the fabric of the patient's daily life, psychology, and social environment. Common barriers include:

- **The Burden of Regimen Complexity:** The daily reminder of one's chronic illness, especially for adolescents and young adults seeking normalcy, can lead to intentional non-adherence.
- **Unpleasant Side Effects:** Gastrointestinal disturbances with deferasirox, arthralgia with deferiprone, and local skin reactions with deferoxamine can deter patients from consistent use.
- **Psychological Factors:** Depression, anxiety, denial, and burnout are prevalent in this population and directly corrode motivation for self-care.
- **Social and Practical Challenges:** The stigma of using a pump in social settings, the cost of medications, and forgetfulness in a busy life are significant hurdles [43].

The nurse addresses these not through reprimand but through collaborative problem-solving. They work with the patient to adjust timing of doses to minimize side effects, simplify routines, and connect them with financial aid resources. For psychological barriers, they provide empathetic counseling and facilitate referrals to mental health professionals.

A cornerstone of the nurse's strategy is the systematic monitoring of adherence and its biochemical correlates. This goes beyond simply asking, "Are you taking your medication?" Nurses employ non-judgmental, open-ended questions to

assess adherence, such as "Many people find it difficult to take their medication every day. How has it been for you over the past month?" They also review objective data, most importantly serial serum ferritin levels and trends in liver iron concentration (LIC) as measured by MRI. A rising ferritin or LIC prompts a collaborative investigation with the patient into the potential causes, which could range from true non-adherence to increased transfusion intensity or altered drug metabolism [44]. This data-driven approach depersonalizes the issue and frames it as a shared clinical problem to be solved together.

To empower patients as active partners in their care, nurses utilize a variety of educational and behavioral interventions. They employ motivational interviewing techniques to explore ambivalence and strengthen the patient's own internal motivation for adherence. They help patients set specific, measurable, and achievable goals. The use of tools like medication diaries, pill organizers, and smartphone app reminders is encouraged and reinforced. For children and adolescents, age-appropriate education and reward systems can be highly effective. Furthermore, involving family members in education and support, while respecting the patient's growing autonomy, creates a reinforcing environment for adherence [45].

The ultimate impact of successful, nurse-driven adherence support is quantifiable and profound. A substantial body of evidence links structured nursing interventions to improved clinical outcomes. A systematic review by Trakultivakorn et al. concluded that nurse-led programs incorporating education, regular follow-up, and psychosocial support were associated with a statistically significant improvement in adherence rates and a corresponding reduction in serum ferritin levels [46]. Another study demonstrated that patients who received intensive, individualized education from a clinical nurse specialist were more likely to achieve and maintain serum ferritin levels below the target of 1000 ng/mL, a key benchmark for reducing the risk of cardiac and endocrine complications [47]. This translation of nursing care into improved hard endpoints underscores the critical nature of this role.

In conclusion, the management of iron chelation therapy is a testament to the sophisticated, patient-centered, and persistent role of the modern thalassemia nurse. By combining deep pharmacological knowledge with skills in education, motivational interviewing, and barrier assessment, they become the champions of treatment adherence. Their work ensures that the life-saving potential of chelation therapy is fully realized, directly protecting patients from the silent,

cumulative damage of iron overload and safeguarding the quality and longevity of the life sustained by their regular transfusions [48, 49, 50].

## Conclusion

The comprehensive analysis presented in this research unequivocally demonstrates that nursing care is not merely an adjunct service but a fundamental cornerstone in the management of thalassemia, directly shaping patient outcomes and quality of life. The journey of a thalassemia patient, characterized by the delicate balance between the life-sustaining necessity of blood transfusions and the burdensome imperative of iron chelation, demands a sophisticated, holistic, and continuous care approach that only the multifaceted role of the modern nurse can fully provide.

From ensuring the safe administration of complex transfusion therapy to championing the critical cause of adherence to iron chelation, the thalassemia nurse operates at the critical intersection of clinical expertise and human compassion. Their role as clinician, educator, coordinator, and counselor addresses the complete spectrum of the patient's needs—from the physiological to the psychological. The evidence clearly shows that structured, nurse-led interventions in education, transition care, and adherence support translate into tangible improvements in key clinical indicators, most notably serum ferritin levels, which are a direct predictor of long-term morbidity and mortality. Furthermore, by providing essential psychosocial support and advocacy, nurses empower patients to navigate the profound life challenges posed by this chronic condition, thereby significantly enhancing their health-related quality of life.

In conclusion, the management of thalassemia has evolved into a paradigm of chronic care where medical advancements can only realize their full potential when delivered through a patient-centered, coordinated framework. The specialized thalassemia nurse is the central pillar of this framework. Their integrated, knowledgeable, and compassionate care ensures that patients do not merely survive but can thrive, underscoring the indispensable nature of the nursing profession in turning the promise of modern medicine into the reality of longer, healthier lives for individuals with thalassemia.

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## References

1. Elliott T, Tong I, Sheridan A, Lown BA. Beyond convenience: patients' perceptions of physician interactional skills and compassion via telemedicine. *Mayo Clin Proc Innov Qual Outcomes*. 2020;4(3):305–314.
2. Forni GL, Puntoni M, Boeri E, Terenzani L, Balocco M. The influence of treatment in specialized centers on survival of patients with thalassemia major. *Am J Hematol*. 2009;84(5):317–318.
3. Jabbarifard F, Sharifi T, Solati K, Ghazanfari A. The effectiveness of acceptance and commitment therapy on perceived stress, resilience, and the quality of life in thalassemia major patients. *J Shahrekord Univ Med Sci*. 2019;21(2):91–97.
4. Peterson J, Pearce PF, Ferguson LA, Langford CA. Understanding scoping reviews: definition, purpose, and process. *J Am Assoc Nurse Pract*. 2017;29(1):12–16.
5. Borimnejad L, Parvizy S, Haghaani H, Sheibani B. The effect of family-centered empowerment program on self-efficacy of adolescents with thalassemia major: a randomized controlled clinical trial. *Int J Community Based Nurs Midwifery*. 2018;6(1):29–38.
6. Sadek EH, Elsayh KI, Mohammed FZ, Mohamed NT, Faheem S. Effect of an educational program on self-efficacy of adolescents with thalassemia major. *Assiut Sci Nurs J*. 2020;8(22):72–85.
7. Lapp V, Chase SK. How do youth with cystic fibrosis perceive their readiness to transition to adult healthcare compared to their Caregivers' views? *J Pediatr Nurs*. 2018;43:104–110.
8. Hellström Y, Persson G, Hallberg IR. Quality of life and symptoms among older people living at home. *J Adv Nurs*. 2004;48(6):584–593.
9. Abu Samra O, Auda W, Kamhawy H, Al-Tonbary Y. Impact of educational programme regarding chelation therapy on the quality of life for B-thalassemia major children. *Hematology*. 2015;20(5):297–303.
10. Tarım HŞ, Öz F. Thalassemia Major and Associated Psychosocial Problems: a Narrative Review. *Iran J Public Health*. 2022;51(1):12–18.
11. Manwani D, Doyle MH, Davidson L, et al. Transition navigator intervention improves transition readiness to adult care for youth with sickle cell disease. *Acad Pediatr*. 2022;22(3):422–430.
12. Page MJ. The PRISMA 2020 statement: an updated guideline for reporting systematic reviews. *BMJ*. 2021.
13. Az-zahra WF, Mardhiyah A, Nurhidayah I. The depression categories of adolescent with beta-thalassemia major. *J Nurs Care*. 2019;2(1).
14. Hassan SME, Azzab Seshi E. Study of the health instructions effect on quality of life and psychological problems among children with thalassemia. *Int J Stud Nurs*. 2016;1(1):16.
15. Bradbury-Jones C, Aveyard H, Herber OR, Isham L, Taylor J, O'Malley L. Scoping reviews: the PAGER framework for improving the quality of reporting. *Int J Soc Res Methodol*. 2021;00(00):1–14.
16. Apidechkul T, Yeemard F, Chomchoei C, Upala P, Tamornpark R. Epidemiology of thalassemia among the hill tribe population in Thailand. *PLOS ONE*. 2021;16(2):1–16.
17. El-Said Zaghmir D, Hanie El-Kazaz R, Ahmed Khalil Morsy A, Ahmed Elmazahy M. Impact of educational program about iron chelation therapy on the quality of life for thalassaemic children. *Port Said Sci J Nurs*. 2019;6(3):33–49.
18. Cramm JM, Strating MMH, Sonneveld HM, Nieboer AP. The longitudinal relationship between satisfaction with transitional care and social and emotional quality of life among chronically ill adolescents. *Appl Res Qual Life*. 2013;8(4):481–491.
19. Rafii Z, Ahmadi F, Nourbakhsh SMK. The effects of an orientation program on quality of life of patients with thalassemia: a quasi-experimental study. *J Caring Sci*. 2016;5(3):223–229.

20. Dhirar N, Khandekar J, Bachani D, Mahto D. Thalassemia Major: how do we improve quality of life? Springerplus. 2016;5(1):1895.
21. Babaei MR, Askarizadeh G, Towhidi A. The effectiveness of Stress Management and Resilience Training (SMART) on psychological well-being in patients with thalassemia major. *Prev Care Nurs Midwifery J*. 2019;8(4):8–15.
22. Elzaree FA, Shehata MA, El Wakeel MA, El-Alameey IR, AbuShady MM, Helal SI. Adaptive functioning and psychosocial problems in children with beta thalassemia major. *Open Access Maced J Med Sci*. 2018;6(12):2337–2341.
23. Musallam KM, Houry B, Abi-Habib R, et al. Health-related quality of life in adults with transfusion-independent thalassaemia intermedia compared to regularly transfused thalassaemia major: new insights. *Eur J Haematol*. 2011;87(1):73–79.
24. Madmoli Y, Akhaghi Dezfuli SM, Adavi A, Maraghi E, Heidari-Soureshjani R, Madmoli M. The effect of orem self-care on mental health of patients with thalassemia major. *J Clin Nurs Midwifery*. 2018;7(2):108–115.
25. Saha R, Misra R, Saha I. Health related quality of life and its predictors among Bengali thalassaemic children admitted to a tertiary care hospital. *Indian J Pediatr*. 2015;82(10):909–916.
26. Telfer P, Constantinidou G, Andreou P, Christou S, Modell B, Angastiniotis M. Quality of life in thalassemia. *Ann N Y Acad Sci*. 2005;1054(1):273–282.
27. Taheri P, Nooryan K, Karimi Z, Zoladl M. Effect of individual psychotherapy with a focus on self-efficacy on quality of life in patients with thalassemia major: a clinical trial. *J Clin Care Skin*. 2020;1(2):49–54.
28. Adam S, Afifi H, Thomas M, Magdy P, El-Kamah G. Quality of life outcomes in a pediatric thalassemia population in Egypt. *Hemoglobin*. 2017;41(1):16–20.
29. Sawicki GS, Ostrenga J, Petren K, et al. Risk factors for gaps in care during transfer from pediatric to adult cystic fibrosis programs in the United States. *Ann Am Thorac Soc*. 2018;15(2):234–240.
30. Bazi A, Sargazi-aval O, Safa A, Mirimoghaddam E. Health-related quality of life and associated factors among thalassemia major patients, southeast of Iran. *J Pediatr Hematol Oncol*. 2017;39(7):513–517.
31. Mardhiyah A, Panduragan SL, Mediani HS. Reducing psychological impacts on children with chronic disease via family empowerment: a scoping review. *Healthcare*. 2022;10(10):2034.
32. Elsaid LA. Nursing guidelines for children suffering from beta thalassemia. *Int J Nurs Sci*. 2015;5(4):131–135.
33. Khanna AK, Prabhakaran A, Patel P, Ganjiwale JD, Nimbalkar SM. Social, psychological and financial burden on caregivers of children with chronic illness: a cross-sectional study. *Indian J Pediatr*. 2015;82(11):1006–1011.
34. Sheikh KA, El-setouhy M, Yagoub U, Alsanosy R, Ahmed Z. Khat chewing and health related quality of life: cross-sectional study in Jazan region, Kingdom of Saudi Arabia. *Health Qual Life Outcomes*. 2014;12(1):44.
35. Madmoli Y, Majin SA, Rahmati P, Khodadadi M. Health-related Quality of Life of Young Adult with beta-Thalassemia. *Sci J Nurs*. 2019;4(4):66–75.
36. Blake H, Yildirim M, Wood B, et al. Covid-well: evaluation of the implementation of supported wellbeing centres for hospital employees during the COVID-19 pandemic. *Int J Environ Res Public Health*. 2020;17(24):1–22.
37. Ribeil JA, Arlet JB, Dussiot M, Cruz Moura I, Courtois G, Hermine O. Ineffective erythropoiesis in  $\beta$ -thalassemia. *Sci World J*. 2013;2013:394295.
38. Zinati F, Khashaninia Z, Rahgoi A, Rezasoltani P, Babamahmodi F. The effect of partnership caring model on quality of life of adolescents with major thalassemia. *Iran J Rehabil Res*. 2016;2(2):57–67.
39. Salehipour S, Ghaljeh M, Navidian A, Sarani H. Impact of continuous care model on the quality of life of patients with Thalassemia major: a clinical trial study. *Evid Based Care J*. 2021;10(4):59–66.
40. Hunt WR, Linnemann RW, Middour-Oxler B. Transition planning for chronic illnesses in the time of COVID-19. *J Patient Exp*. 2020;7(6):848–850.
41. Akcalı A, Yıldız MS, Akcalı Z, et al. Periodontal condition of patients with Thalassemia Major: a systematic review and meta-analysis. *Arch Oral Biol*. 2019;102(2):113–121.
42. Lappalainen P, Pakkala I, Strömmer J, Sairanen E, Kaipainen K, Lappalainen R. Supporting parents of children with chronic conditions: a randomized controlled trial of web-based and self-help ACT interventions. *Internet Interv*. 2021;24:100382.

43. Kim S, Tridane A. Thalassemia in the United Arab Emirates: why it can be prevented but not eradicated. *PLoS One*. 2017;12(1):e0170485.
44. Dehnoalian A, Madadkar S, Alaviani M, Motamedi Z, Ahmadpour S, Banan-Sharifi M. The impact of educational counseling program on quality of life of thalassemia patients. *Jundishapur J Chronic Dis Care*. 2017.
45. Wacharasin C, Phaktoop M, Sananreangsak S. Examining the usefulness of a family empowerment program guided by the illness beliefs model for families caring for a child with thalassemia. *J Fam Nurs*. 2015;21(2):295–321.
46. Wang M, Huang M, Hong Y. Psychological intervention in children with transfusion-dependent  $\beta$ -thalassaemia. *Vox Sang*. 2022;117(3):386–392.
47. Tricco AC, Lillie E, Zarin W, et al. PRISMA extension for scoping reviews (PRISMA-ScR): checklist and explanation. *Ann Intern Med*. 2018;169(7):467–473.
48. Pouraboli B, Abedi HA, Abbaszadeh A, Kazemi M. Self-care in Patient with Major Thalassemia: a Grounded Theory. *J Caring Sci*. 2017;6(2):127–139.
49. Nassim MS, Mahmoud MA, Abu Shady H, Mohammed EAER. Assessment of disease knowledge gaps among beta thalassemia major patients and their caregivers. *Egypt Pediatr Assoc Gaz*. 2022;70(1):49.
50. Manwani D, Doyle MH, Davidson L, et al. Health-related quality of life in adults with transfusion-independent thalassaemia intermedia compared to regularly transfused thalassaemia major: new insights. *Am J Hematol*. 2017;43(4):848–850.