

THE NUTRITIONAL STATUS OF PATIENTS WITH BETA THALASSEMIA MAJOR: A SCOPING REVIEW

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ABSTRACT

Objective: This study aimed to review and summarize the existing evidence on the nutritional status of patients with beta thalassemia major.

Methods: A scoping review was conducted using the Medline database via PubMed. Seven studies from six countries, involving 836 participants, met the inclusion criteria. Data were extracted on anthropometric measurements, body composition, laboratory indicators, and dietary intake.

Results: The findings indicated a high prevalence of malnutrition, particularly among children with beta thalassemia major. Anthropometric data showed significantly lower weight, height, and body mass index compared to healthy individuals. Body composition analysis revealed lower fat percentages and higher muscle mass in children, while adults exhibited decreased lean mass and increased fat mass, with 64.2% classified as obese with sarcopenia. Laboratory results showed deficiencies in serum vitamin D and cholesterol, though calcium and phosphorus levels remained normal. Dietary intake assessments revealed inadequate energy and nutrient intake, particularly for macronutrients and key micronutrients such as calcium, phosphorus, and zinc.

Conclusion: Patients with beta-thalassemia major are at high risk of malnutrition, which is associated with both disease and treatment factors. Routine nutritional assessments and tailored interventions are critical for improving the nutritional status and overall health outcomes of this vulnerable population.

Keywords: nutritional status, patient, Beta Thalassemia major, scoping review, malnutrition.

1. BACKGROUND

Beta-thalassemia is an inherited disorder caused by a mutation in the beta-globin gene, leading to a reduction in the production of the beta-globin chain of hemoglobin. The severity of the disease varies, ranging from minor (with no clinical symptoms), to intermedia (patients showing symptoms but not requiring regular blood transfusions), and major (requiring frequent blood transfusions). Blood transfusion is the main treatment for patients with beta-thalassemia major, but it can lead to iron overload, which causes serious complications in the liver, kidneys, and endocrine system.

Due to the nature of the disease and the side effects of the treatment, patients with beta-thalassemia major often experience nutritional deficiencies. Studies have shown that these patients are frequently deficient in essential amino acids, vitamins, and minerals compared

to healthy individuals [1, 2]. This is primarily due to reduced appetite and fatigue from anemia, along with impaired digestion and nutrient absorption [3].

Maintaining good nutritional status is critical for patients with beta-thalassemia major as it helps enhance immune function, supports ongoing treatment, and reduces the risk of complications [4]. However, there is currently very little published data on the nutritional status of these patients. Therefore, we conducted this study with the objective of reviewing the existing evidence on the nutritional status of this patient group.

2. METHODS

2.1. Search strategy and selection criteria

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This scoping review explores the nutritional status of patients with beta-thalassemia major published until 30 May 2024. We included studies published in peer-reviewed journals in English. Exclusion criteria were laboratory studies, animal studies, and non-original research articles (such as letters to the editor, viewpoints, and narrative reviews).

Our literature search was conducted using the Medline database via PubMed, applying the following search terms: "nutritional Status"[MeSH Terms] AND ("beta-Thalassemia"[MeSH Terms] OR "beta-Thalassemia major"[All Fields]). The results were imported into Rayyan software, where two independent researchers (HTNA and DTTT) screened the titles and abstracts for relevance. Full texts of the selected studies were then retrieved and evaluated based on pre-established eligibility criteria. Any discrepancies during the screening process were resolved through discussion, with a third researcher (BTMA) consulted when necessary. Reasons for exclusion were documented, and a PRISMA flowchart was developed to illustrate the study selection process.

2.2. Data extraction

Data were extracted by one researcher (HTNA) using a standardized extraction form. Extracted data included

general study information (authors, journal, publication date, and year of data collection), study design, participant details (country of patient enrollment and number of participants), and the nutritional status of beta-thalassemia patients (anthropometric methods, body composition measurements, laboratory indices, and dietary intake).

2.3. Data analysis

The extracted data were entered into a Microsoft Excel database. Variables related to general study information and nutritional status were systematically described for each study.

3. RESULTS

3.1. Selection process

A total of 34 search results were retrieved from MEDLINE via PUBMED. After screening the titles and abstracts for relevance, 14 articles were excluded. During the full-text review for eligibility, an additional 13 articles were excluded, primarily due to incorrect study designs. Ultimately, 7 studies were included, and their data were extracted for analysis.

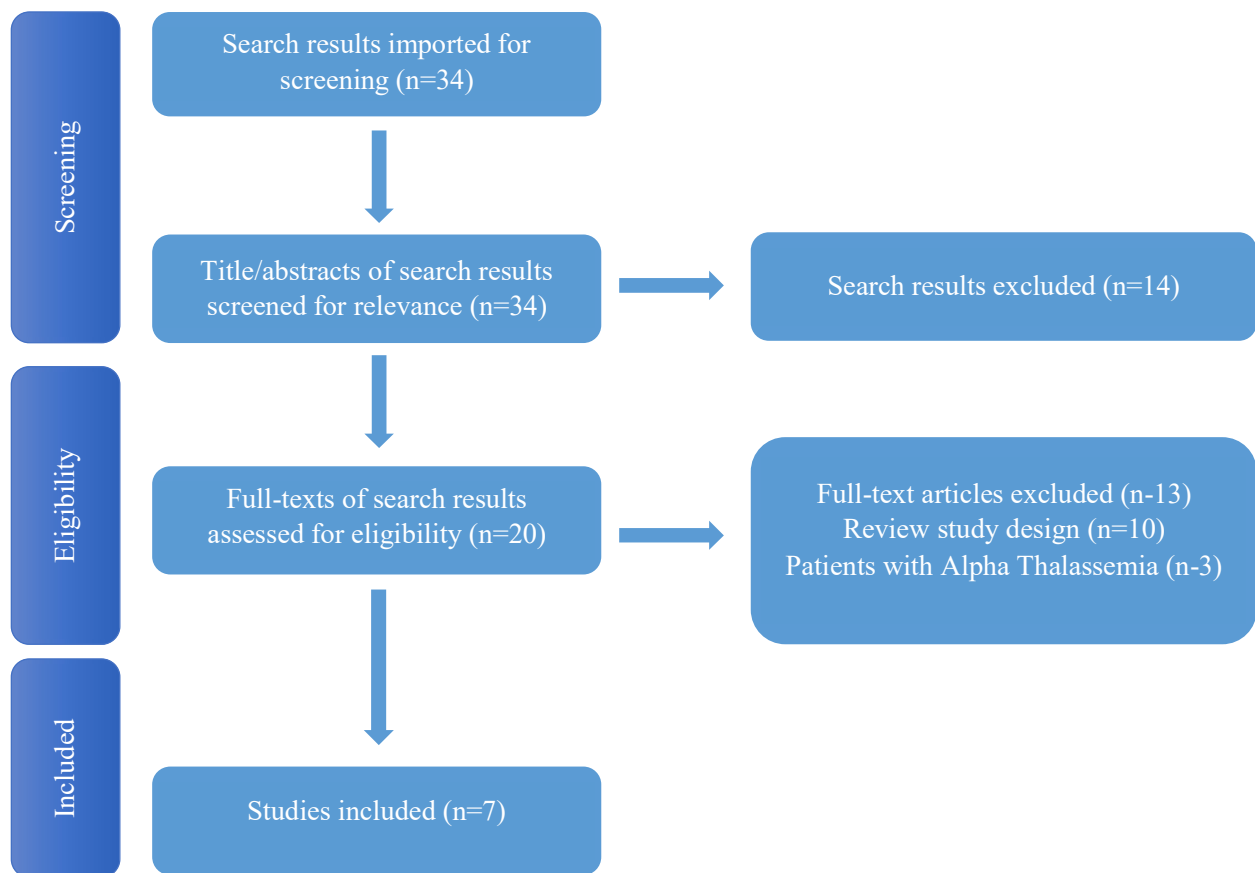


Figure 1. Selection of studies on nutritional status of patients with beta-thalassemia major

3.2. General characteristics of the articles

The publication dates of the seven selected articles range from 2003 to 2023, covering research conducted in six countries: Egypt (two studies), Iran, Thailand, Sri Lanka, Greece, and Turkey. In terms of study design, four articles employed cross-sectional designs, one was a case-control study, one was an intervention study without a control group, and one was a retrospective cohort study. The participants included both children (in four studies) and adults (in three studies), with a pooled sample size of 836 patients (Median: 89, Interquartile Range: 67–200).

3.3. Nutritional status of patients with beta-thalassemia major

The nutritional status of patients was evaluated using four primary indicators: anthropometric measurements, body composition analysis, laboratory assessments, and dietary intake.

Anthropometric Measurements

Most studies indicated that patients with beta-thalassemia major experienced malnutrition, particularly among children. For instance, Naghmed et al. (2013) conducted a study in Iran on 140 children with beta-thalassemia major, finding that 33.6% were malnourished (defined as a z-score < -2SD). In contrast, Irene et al. (2022) examined 67 adults in Greece and found that 7.5% of the participants were malnourished, as assessed by BMI and the MUST nutrition screening tool [5]. Across the studies, patients with beta-thalassemia major consistently had lower weight, height, and body mass index compared to healthy individuals of the same age and gender [6, 7].

Body composition

Four of the seven included studies assessed body composition. Children with beta-thalassemia major were found to have a significantly lower percentage of body fat and a higher percentage of muscle compared to healthy children of the same age and gender [6]. Despite this, bone mineral density and bone mineral content were within the normal range for these children [6, 7]. In contrast, adult patients displayed different trends, with 86.8% exhibiting decreased lean mass and 74.6% showing increased fat mass. Notably, up to 64.2% of adult patients were classified as obese with sarcopenia [6, 8].

Laboratory Indicators

The laboratory indicators used to assess the nutritional status of patients with beta-thalassemia major varied across studies. Serum vitamin D levels were measured in three studies, while total cholesterol, calcium, and phosphorus levels were assessed in two studies each. The results showed that both total cholesterol and serum vitamin D levels were significantly lower than normal [5-8]. However, calcium and phosphorus levels

in these patients remained within normal ranges [7].

Dietary Intake

Dietary assessments were conducted in two of the seven studies, both involving children. These studies indicated that the patients' energy intake did not meet their bodies' energy requirements [6]. Furthermore, the intake of macronutrients (carbohydrates, proteins, and fats) and micronutrients (calcium, phosphorus, and zinc) was consistently lower than the recommended levels [6, 7].

4. DISCUSSION

This study provides a review of the existing literature on the nutritional status of patients with beta-thalassemia major, highlighting several key areas of concern. The findings indicate that malnutrition is a prevalent issue in this population, particularly among children, and is manifested across multiple nutritional indicators, including anthropometric measurements, body composition, laboratory indices, and dietary intake.

One of the most consistent findings across the studies reviewed was the high prevalence of malnutrition among patients with beta-thalassemia major, especially children. Malnutrition was defined by low z-scores, body mass index (BMI), and other anthropometric indicators. In the studies by Naghmed et al. (2013) and Irene et al. (2022), children and adults with beta-thalassemia major showed lower weight, height, and BMI compared to healthy individuals [5]. This highlights the chronic nature of nutritional challenges in this patient population. Given the role of proper nutrition in supporting immune function and mitigating the effects of anemia and other complications, the findings emphasize the urgent need for targeted nutritional interventions.

The body composition analysis further underscored disparities in nutritional status between beta-thalassemia patients and their healthy peers. Children with beta-thalassemia major exhibited lower body fat percentages and higher muscle mass, while adults, particularly those with longer disease duration, tended to exhibit higher fat mass and decreased lean mass [6]. Notably, up to 64.2% of adult patients were classified as obese with sarcopenia [9]. This could be attributed to the combination of reduced physical activity and the long-term effects of iron overload and chronic disease management. These body composition shifts highlight the need for a tailored approach to nutritional and physical therapy that addresses both the immediate and long-term nutritional needs of patients with beta-thalassemia major.

Laboratory indicators provided additional insight into the biochemical aspects of malnutrition in this population. Low serum vitamin D and cholesterol



levels were common across several studies, indicating deficiencies that could have significant implications for bone health and overall metabolic function [5-8]. The finding that calcium and phosphorus levels remained within normal ranges may suggest some compensation in bone metabolism, but the overall picture points to the need for regular monitoring of these biochemical markers to prevent deficiencies from leading to more severe complications such as osteoporosis and cardiovascular disease [7].

Dietary intake data, although limited, revealed that the energy and nutrient intake of patients, particularly children, was consistently lower than recommended levels. The insufficient intake of both macronutrients (carbohydrates, proteins, and fats) and micronutrients (calcium, phosphorus, zinc) further underscores the nutritional inadequacies faced by these patients. This likely results from a combination of factors, including reduced appetite, fatigue, and impaired digestion, as previously documented in beta-thalassemia patients. The observed dietary deficits call for comprehensive nutritional assessments and interventions aimed at meeting the specific needs of this patient group [6].

Overall, this review highlights the multidimensional nature of malnutrition in patients with beta-thalassemia major. The findings stress the importance of implementing routine nutritional assessments as part of the management of these patients, with particular attention to both children and adults. Furthermore, addressing the challenges of iron overload, muscle mass loss, and micronutrient deficiencies should be integrated into the long-term care plans of patients with beta-thalassemia major. Interventions focusing on improving dietary intake, monitoring body composition, and correcting laboratory abnormalities may have a significant impact on improving quality of life and reducing complications related to malnutrition.

Limitations

Despite the important insights provided by this review, several limitations should be acknowledged. First, the limited number of studies restricts the generalizability of the findings. Moreover, the heterogeneity of study designs and methods, including the varied definitions of malnutrition and different approaches to nutritional assessments, complicates the synthesis of results. Additionally, the studies reviewed largely focused on cross-sectional data, limiting the ability to draw conclusions about the long-term nutritional trajectory of these patients. Future research should aim to address these gaps by including longitudinal studies with standardized nutritional assessments across different age groups and regions.

5. CONCLUSION

This study underscores the critical importance of addressing malnutrition in patients with beta-thalassemia major. The consistent finding of poor nutritional status across anthropometric, body composition, laboratory, and dietary assessments suggests that comprehensive nutritional management should be a central component of beta-thalassemia care. Implementing tailored nutritional interventions and monitoring strategies could significantly improve patient outcomes and reduce the risk of complications associated with malnutrition in this vulnerable population.

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