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Quality of Life among Paediatric Patients with Transfusion-Dependent Beta-Thalassemia Major in Sabah, Malaysia

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ABSTRACT

Background: Thalassaemia is a significant health concern in Malaysia, particularly in Sabah, where prevalence rates are high. This study aims to determine the quality of life (QOL) of paediatric patients with β-thalassaemia major (β-TM) in Sabah and to identify factors associated with the QOL. **Methods:** A cross-sectional study was conducted at two hospitals between November 2018 and April 2019, including 115 transfusion-dependent β-TM patients aged 8 to 18 years. The Malay version 4.0 of the PedsQLTM questionnaire was used to assess QOL. Paired t-test was used to compare the QOL scores between patients and their parents' proxy reports. Linear regression analysis was performed to identify factors influencing QOL scores. **Results:** The mean total QOL summary score was 76.15 (SD 14.5), with the lowest mean score observed in school functioning (64.1). Only the school functioning score was significantly lower (p-value = 0.02) in the parents' proxy report compared to the patients' self-report. In multivariable analysis, parental marital status (adjusted $\beta = -10.55$, 95% CI −19.38, −1.73, $p = 0.02$) and the usage of single iron chelation therapy or monotherapy (adjusted $\beta = 10.02$, 95% CI 4.05, 16.00, $p = 0.001$) were significantly associated with the lower total QOL summary score. **Conclusion:** Thalassaemia negatively impacts patient's QOL, especially in the school functioning. This study recommended considering QOL as an important factor in the management of transfusion-dependent thalassaemia patients, which includes providing out-of-hours thalassaemia treatment services and implementing nurse-led programmes that offer social support for patients and families.

Keywords: Beta-thalassemia Major; Paediatric; PedsQL; Quality of Life; Health

INTRODUCTION

Thalassaemia major patients require long-term blood transfusions and iron chelation therapy, which can lead to potential complications such as bloodborne infections, iron overload, growth retardation, splenomegaly, and delayed sexual development (Nienhuis & Nathan, 2012). In Malaysia, thalassemia is the most common single-gene disorder, with approximately 4.5% of the population being carriers of β-thalassaemia (George, 2001). As of November 2018, the Malaysian Thalassaemia Registry reported 8,681 registered thalassaemia patients, with 4,529 receiving regular transfusions (Ibrahim *et al*., 2020). This condition remains a significant public health concern in Malaysia, particularly in Sabah, which has the highest reported prevalence of thalassaemia.

Nurses play a vital role in the multidisciplinary management of thalassaemia by providing direct care during transfusions, monitoring for transfusion-related complications, educating patients and families about chelation therapy adherence, and ensuring infection control practices. They are instrumental in early detection of adverse effects through routine assessments and contribute to care coordination by liaising with physicians, dietitians, and pharmacists. Furthermore, nurses often provide emotional support, counselling, and patient advocacy, which are crucial in managing the psychological and social stressors faced by patients and their families.

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Living with thalassaemia can have a significant impact on patients' lives, as they often require frequent hospital visits for transfusions and face complications from iron chelation therapy, despite advancements in clinical management (Bakhshi *et al*., 2017; Olivieri & Brittenham, 2013). These challenges can lead to psychological, emotional, and social difficulties for both patients and their families, including regular school absenteeism, limited family activities, coping struggles, mental health issues, and financial burdens (Ishfaq *et al.*, 2015; Tarim & Oz, 2022). Therefore, assessing the Quality of life (QOL) of thalassaemia patients is crucial not only to understand their overall well-being but also to develop targeted nursing interventions that can enhance their physical, emotional, and psychosocial health. A comprehensive evaluation allows nurses to implement personalised care strategies that improve holistic health outcomes and support long-term disease management.

QOL is a multifaceted concept that entails a subjective assessment of overall well-being, including psychological, physical and social functioning (Revicki *et al*., 2014). In Malaysia, only four studies have examined the QOL of children with thalassaemia (Ismail *et al*., 2013; Sazlina *et al*., 2015; Shafie *et al*., 2020). Three of these studies focused solely on the central states of West Malaysia, while the most recent and largest study included 368 patients from different regions across Malaysia, including East Malaysia. However, there is a lack of comprehensive QOL studies specifically addressing the Sabah population. Sabah has diverse indigenous populations and higher poverty rates compared to West Malaysia, which may expose thalassaemia patients to unique challenges affecting their QOL. These experiences remain underexplored, highlighting the need for further research. This study aims to determine the QOL scores of paediatric patients with β-thalassaemia major in the Sabah population and identify the factors associated with their QOL.

METHODOLOGY

Research Design and Study Participants

A cross-sectional study was conducted at two Thalassaemia Treatment Centres (TTC) in Sabah, Malaysia: Hospital Wanita dan Kanak-Kanak Likas (HWKKS) and Hospital Kota Belud (HKB). Data collection took place between November 2018 and April 2019. Patients attending the TTC for their routine review were purposively selected based on their appointment schedule, as simple random sampling was not feasible due to the limited sample. This approach aligns with established methodologies in rare disease research, where purposive sampling ensures inclusion of information-rich cases that meet specific clinical criteria (Patton, 2015). The study included transfusion-dependent *β*-thalassaemia major patients aged 8 to 18 years old. Exclusion criteria included patients who were non-transfusion dependent, illiterate, in poor health, unwilling to participate, or had undergone bone marrow transplants. Parents accompanying their thalassaemia child were selected as participants (as proxy reports). However, for older patients who were capable of self-reporting and not accompanied by a parent, proxy reports were not used.

The sample size was determined using a single mean formula. Based on a previous study, the Standard Deviation (SD) of the QOL summary score among paediatric patients with *β*-thalassaemia in Malaysia was 13.1 (Sazlina *et al*., 2015). This study aimed to estimate the mean QOL summary score among paediatric patients in Sabah within a 2.5 points margin of error and a 95% confidence interval (CI). Considering a 20% non-response rate (Xu *et al*., 2018), the minimum sample size was 126. Additionally, the sample size was also calculated for the second objective, with the final sample size chosen based on the highest sample from the calculations. However, due to exclusion criteria and an inadequate number of eligible participants, only 115 participants were included in this study.

Study Instrument

A data collection form comprising three sections was utilised in this study. Section A recorded patients' demographic information (age, gender, ethnicity, and residential town/district) and socioeconomic information (schooling status, education level, total monthly household income, parents' marital status and financial aid status). Section B documented parents' information, detailing the number of children in the family, the number of children with thalassaemia carrier status, and the number of children with *β*-thalassaemia major. *Section C* documented patients' clinical information (age of diagnosis, age of transfusion, blood type, average pre-transfusion haemoglobin [Hb] level, serum ferritin level, types of iron chelation therapy, presence of splenomegaly, presence of endocrine complications, and presence of co-morbidities) obtained from patients' case notes.

QOL was assessed using the self-reported Malay version 4.0 of the Paediatric Quality of Life InventoryTM (PedsQLTM) questionnaire, obtained from the MAPI Research Institute, France (Varni *et al*., 2001). The questionnaire was administered to both patients and their parents. The PedsQLTM instrument is a validated and reliable tool for measuring the health-related QOL of paediatric populations with acute or chronic health conditions. The self-reported PedsQLTM demonstrated high internal consistency approaching 0.9 and has been confirmed as reliable based on previous studies using the Malay version (Ismail *et al*., 2013; Sazlina *et al*., 2015). The PedsQLTM questionnaire consists of 23 items and encompasses four domains: physical functioning (8 items), emotional functioning (5 items), social functioning (5 items) and school functioning (5 items). The questionnaire assessed physical, emotional, social, and schooling function-related challenges over the past month. Responses were recorded using a five-point rating scale, where 0 indicated 'never a problem' and 4 indicated 'almost always a problem'. Total scores were calculated, along with two summary scores: physical health and psychosocial health (averaging emotional, social, and school functioning). All items were inversely scored and converted to a 0 - 100 scale, where higher scores indicate better QOL.

Data Collection Method

The study was explained to eligible patients and their parents, who were then invited to participate voluntarily. Before participation, assent from the patients and informed consent from their parents were obtained. Each patient and their parent were given the PedsQLTM 4.0 questionnaire and instructed to complete it individually on the same day. Sociodemographic data, parents' information, and clinical data were obtained from patients' case notes. If any information was missing, the researcher conducted interviews with the participants to collect the necessary details. The researcher was present while the participants completed the questionnaire to assist with any questions.

Statistical Analyses

The data was analysed using IBM SPSS version 24.0. Descriptive statistics were used to summarise the data. Categorical variables were presented as percentages and frequencies. Numerical variables were reported as mean and SD. For skewed numerical variables, the median and Interquartile Range (IQR) were reported. A paired *t*-test was used to compare the QOL scores between patients and their parents. Normality assumption was checked and met before applying the test. Linear regression analysis was performed to identify factors associated with physical health score, psychosocial health score, and total summary score. Assumptions such as linearity, independence, homoscedasticity and normality of residuals were checked to ensure robustness of the results. The simple linear regression analysis included important factors for QOL as reported in previous studies, as well as other potential factors that could affect patients' QOL. These factors were age, age of diagnosis, gender, ethnicity, education level, residential area, number of siblings, presence of siblings with β-thalassaemia major, parents' marital status, total monthly household income, receipt of financial aid, average pre-transfusion Hb level, types of iron chelation therapy, serum ferritin level, and growth and development percentile. Certain variables were excluded to avoid multicollinearity and ensure accurate estimation. Specifically, age at transfusion was highly correlated with age at diagnosis. Splenectomy and endocrine complications were excluded due to the limited number of patients with these conditions. In the multivariable analysis, variableswith a *p*-value below 0.25 in the univariable analysis were selected, consistent with the recommended threshold for variable inclusion in model building (Bursac *et al*., 2008). Both forward and backward (likelihood ratio) methods were used to identify variables, as these approaches provide robust model selection while controlling for overfitting (Hosmer *et al*., 2013). The level of significance for the study was set at 0.05 (two-tailed).

Ethical Consideration

The researchers obtained ethical clearance from the Research and Ethics Committee of Universiti Sains Malaysia, with reference number USM/JEPeM/18010008 on 15th March 2018. Additionally, the study received approval from the Ministry of Health, Malaysia and the Medical Research and Ethics Committee (MREC), Malaysia with reference number NMRR ID: NMRR-18-218-39524 (IIR) on 1st June 2018.

RESULTS

Sociodemographic and Clinical Characteristics of Thalassemia Patients

This study included 115 transfusion-dependent β-thalassaemia major patients receiving treatment at two

TTCs in Sabah. Among them, 97 patients were from HWKKS, while 18 were from HKB. Initially, 132 patients were identified as receiving treatment at these centres. However, 15 patients were excluded from the study due to illiteracy (school dropout) and having neurocognitive disabilities such as autism and ADHD, which affected their ability to read and follow instructions. Furthermore, two patients at HWKKS who had undergone a bone marrow transplant were also excluded.

Table 1 presents the sociodemographic characteristics of the patients. The mean age was 12 years (SD 2.8), with the majority (58.3%) aged between 8 and 12 years. Their gender distribution was equal, with an equal distribution of male and female patients. Most patients (67.8%) were of Kadazandusun Murut (KDM) ethnicity and lived in Kota Kinabalu. Nearly all (97.4%) were still attending school, with the majority in primary school (58.3%). Regarding family background, 35.7% of patients had siblings with thalassaemia major, while 60.9% were the first in their families to be diagnosed. Approximately 10% had divorced parents. The median total monthly household income was 1,500 Malaysian Ringgit (MYR), with 85.2% coming from lower-income families earning less than MYR 4360 per month (Khazanah Research Institute, 2018). Only 32.2% of patients received financial aid from the government.

Table 1: Sociodemographic Characteristics of Patients (n=115)

Sociodemographic Characteristics	Frequency (Percentage)
Age Group (years)	12.0 (2.8) [a]
8 – 12	67 (58.3)
13 – 18	48 (41.7)
Gender	
Female	56 (48.7)
Male	59 (51.3)
Ethnicity	
Non-KDM	36 (31.3)
KDM	78 (67.8)
Missing Information	1 (0.9)
Schooling Status	
Still attending school	112 (97.4)
Quit school	3 (2.6)
Education level	
Primary school	67 (58.3)
Secondary school	48 (41.7)
Reside in Capital City (Kota Kinabalu)	
No	51 (44.3)
Yes	64 (55.7)
Number of Siblings	4 (3 – 5) [b]
Have Siblings with Beta - Thalassemia Major	
No	67 (58.3)
Yes	41 (35.7)
Missing Information	7 (6.1)
First Affected Child in the Family	
No	21 (18.3)
Yes	70 (60.9)
Missing Information	24 (20.9)
Parents' Marital Status	
Not divorced	79 (68.7)
Divorced	12 (10.4)
Missing Information	24 (20.9)
Total Monthly Household Income (Malaysian Ringgit) [c]	1500 (945 – 3000) [b]
< 4360 (B40 or lower-class income group)	98 (85.2)
4360 – 9619 (M40 or middle-class income group)	9 (7.8)
> 9619 (T20 or upper-class income group)	3 (2.6)
Missing Information	5 (4.3)
Receiving Financial Aids	
No	78 (67.8)
Yes	37 (32.2)

a Mean (Standard deviation) b Median (Interquartile range) KDM: Kadazandusun Murut
c Categorization of household income groups is based on the 2018 report from Khazanah Research Institute

Table 2 presents the clinical characteristics of the patients. The majority (49.6%) had blood type O and 46.1% of patients fell below the third percentile in growth and development. About half of the patients received monotherapy iron chelation, with Deferasirox being the most common treatment (27%). The mean pre-transfusion Hb level was 8.6 g/dL (SD 1.0). The median serum ferritin level was 1766.1 ng/mL (IQR 1065.3–4161.8 ng/mL). Only 1.7% of patients underwent splenectomy and 3.5% experienced endocrine complications such as hypogonadism.

Table 2: Clinical Characteristics of Patients (n=115)

Clinical Characteristics	Frequency (Percentage)
Age of Diagnosis (Years)	1 (0.7 – 2.8) [a]
Age of First Transfusion (Years)	1 (0.8 – 2.8) [a]
Blood Type	
A	23 (20.0)
B	11 (9.6)
AB	2 (1.7)
O	57 (49.6)
Missing	22 (19.1)
Growth and Development Percentile	
< 3rd percentile	53 (46.1)
3rd – 25th percentile	40 (34.8)
Missing	22 (19.1)
Types of Iron Chelation Therapy	
Monotherapy	58 (50.4)
Combination therapy	49 (42.6)
Missing	8 (7.0)
Types of Iron Chelation Therapy - (Chelation Regimen)	
Deferoxamine (Desferal only)	6 (5.2)
Deferiprone only	21 (18.3)
Deferasirox only	31 (27.0)
Deferoxamine (Desferal) + Deferiprone	27 (23.5)
Deferoxamine (Desferal)+ Deferasirox	12 (10.4)
Deferiprone + Deferasirox	10 (8.7)
Missing	8 (7.0)
Average Pre-transfusion Hb Level	8.6 (1.0) [b]
< 9 g/dL	62 (53.9)
≥ 9 g/dL	49 (42.6)
Missing	4 (3.5)
Serum Ferritin Level	1766.1.00 (1065.3 – 4161.8) [a]
< 2500	64 (55.7)
≥ 2500	42 (36.5)
Clinical Characteristics	
Missing	9 (7.8)
Splenectomy	
No	113 (98.3)
Yes	2 (1.7)
Presence of Endocrine Complications	
No	111 (96.5)
Yes	4 (3.5)
Presence of Comorbidity	
No	115 (100.0)
Yes	0 (0.0)

a Median (Interquartile range) bMean (Standard deviation) Hb: Haemoglobin

PedsQL QOL scores from thalassemia patients and their parents

The QOL scores obtained from the PedsQLTM questionnaire are presented in Table 3. The mean total summary scores were 76.15 (SD 14.5) for patients' self-reports and 72.62 (SD 14.86) for parents' proxy reports. There was no significant difference between the patients' self-reported scores and their parents' proxy report in the overall total summary score (p = 0.09). Among the QOL domains, social functioning had the highest mean score (84.17, SD 16.69). School functioning had the lowest mean score (64.09, SD 17.8). Notably, the parents' proxy report showed a significantly lower score for school functioning compared to the patients' self-report (p = 0.02).

Table 3: PedsQL Quality of Life Scores from Patients with Thalassemia and their Parents

Health Related Qualify of Life Score	Patients with Thalassemia (n=115)	Parents of Patients with Thalassemia (n=83)	Mean Difference (95% CI)[a]	*P*-value [a]
	Mean (SD)	Mean (SD)		
Physical functioning	78.10 (15.71)	74.69 (19.60)	3.09 (-1.00, 7.19)	0.14
Psychosocial functioning	75.12 (15.45)	71.51 (14.47)	2.70 (-0.67, 6.08)	0.12
Emotional functioning	77.09 (20.36)	73.86 (18.66)	0.78 (-3.53, 5.09)	0.72
Social functioning	84.17 (16.69)	81.81 (16.54)	1.51 (-2.33, 5.34)	0.44
School functioning	64.09 (17.80)	58.88 (19.41)	5.82 (0.80, 10.85)	**0.02**
Total summary score	76.15 (14.50)	72.62 (14.86)	2.84 (-0.43, 6.11)	0.09

[a]*Paired t-test was performed on 83 pairs of patients and their parents; CI: Confidence Interval*

Factors associated with QOL scores

(i) Physical health summary score

Six variables with a p-value below 0.25 in the univariable analysis were selected for multivariable analysis: number of siblings, having siblings with thalassaemia major, age of diagnosis, average pre-transfusion Hb level, types of iron chelation therapy, and serum ferritin level. In the multivariable analysis, only the serum ferritin level remained significantly associated with the physical health summary score (Table 4). A serum ferritin level of more than 2500 ng/mL was associated with a higher physical health score (adjusted β=9.49, 95% CI 2.76, 16.21).

(ii) Psychosocial health summary score

Seven variables with a p-value below 0.25 in the univariable analysis were selected for multivariable analysis: having siblings with thalassaemia major, age of diagnosis, parents' marital status, income group, average pre-transfusion Hb level, types of iron chelation therapy, and serum ferritin level. In the multivariable analysis, two variables (parents' marital status and types of iron chelation therapy) were significantly associated with the psychosocial health summary score (Table 4). Patients with divorced parents had a significantly lower psychosocial score compared to those with non-divorced parents (adjusted β = −13.81, 95% CI −22.99, −4.62). Patients receiving combination therapy had a significantly higher psychosocial score compared to those on monotherapy (adjusted β = 9.45, 95% CI 2.86, 16.04).

(iii) QOL total summary score

Seven variables with a p-value below 0.25 in the univariable analysis were selected for multivariable analysis: having siblings with thalassaemia major, age of diagnosis, parents' marital status, income group, average pre-transfusion Hb level, types of iron chelation therapy, and serum ferritin level. In the multivariable analysis, two variables (parents' marital status and types of iron chelation therapy) showed significant association with the total summary score (Table 4). Patients with divorced parents had a lower total summary score compared to those with non-divorced parents (adjusted β = −10.55, 95% CI −14.38, −1.73). Patients receiving combination therapy had a higher total summary score than those on monotherapy (adjusted β = 10.02, 95% CI 4.05, 16.00).

Table 4: Univariable and Multivariable Linear Regression Analyses of Factors Associated with Patients' Physical Health, Psychosocial Health, and Total Summary Scores (n=115)

Factors	Physical Health				Psychosocial Health				Total Summary Score			
	Univariable Linear Regression		Multivariable Linear Regression		Univariable Linear Regression		Multivariable Linear Regression		Univariable Linear Regression		Multivariable Linear Regression	
	β (95% CI)	*P*-value	*β* (95% CI)	*P*-value	*β* (95% CI)	*P*-value	*β* (95% CI)	*P*-value	*β* (95% CI)	*P*-value	*β* (95% CI)	*P*-value
Number of Siblings	-1.24 (-2.99, 5.18)	0.165	-1.77 (-3.60, 0.06)	0.058	-0.19 (-1.91, 1.53)	0.828	-	-	-0.55 (-2.17, 1.07)	0.500	-	-
Have Siblings with Beta-Thalassemia Major												
No	Ref				Ref				Ref			
Yes	5.17 (-0.99, 11.32)	0.099	-	-	4.45 (-1.59, 10.49)	0.147	-	-	4.70 (-0.96, 10.36)	0.102	-	-
Parents' Marital Status												
Non-Divorced	Ref				Ref		Ref		Ref		Ref	
Divorced	-2.87 (-12.50, 6.75)	0.555	-	-	-10.6 (-19.95, -1.29)	0.026	-13.81 (-22.99, -4.62)	**0.004**	-7.93 (-16.87, 1.02)	0.082	-10.55 (-14.38, -1.73)	**0.02**
Average Pre-Transfusion Hb Level												
< 9 g/dL	Ref				Ref				Ref			
≥ 9 g/dL	4.04 (-1.92, 10.0)	0.182	-	-	4.39 (-1.38, 10.16)	0.134	-	-	4.27 (-1.17, 9.71)	0.123	-	-
Types of Iron Chelation Therapy												
Monotherapy	Ref				Ref		Ref		Ref		Ref	
Combination Therapy	3.94 (-2.18, -10.05)	0.204	-	-	6.42 (0.59, 12.54)	0.031	9.45 (2.86, 16.04)	**0.006**	5.56 (0.04, 11.08)	0.048	10.02 (4.05, 16.00)	**0.001**
Serum Ferritin Level												
< 2500 ng/mL	Ref		Ref		Ref				Ref			
≥ 2500 ng/mL	6.67 (0.46, 12.89)	0.036	9.49 (2.76, 16.21)	**0.006**	7.34 (1.35, 13.38)	0.017	-	-	7.13 (1.47, 12.79)	0.014	-	-

β: Regression coefficient ; CI: Confidence Interval ; Ref: Reference group Hb: Haemoglobin; Multicollinearity was checked and not found

DISCUSSION

This study found that parents' assessments of their children's overall QOL score closely aligned with the patients' self-assessment, indicating that parent-proxy reports can serve as a reliable measure. This alignment may reflect the close parent-child relationships and strong family support systems characteristic of our study population, which likely enhanced parental awareness of their child's well-being (Ismail *et al*., 2016). The chronic nature of transfusion-dependent *β*-thalassaemia major, requiring lifelong treatment initiated in early childhood, may have further contributed to parents' accurate perceptions, as they develop extensive experience managing their child's health needs over time. However, our findings contrast with recent research by Youssry *et al.* (2023), who reported that parents systematically underestimated their children's QOL compared to self-reports. These discrepancies may be explained by cultural differences in illness perception or variations in disease severity across study populations. Notably, we observed that parents in our study rated school functioning significantly lower than children's self-reports, a pattern consistent with previous findings by Sinlapamongkolkul and Surapolchai (2020). This discrepancy could be due to parents' limited exposure to their child's school environment, making it difficult for them to fully assess their child's academic coping mechanisms. While children may adapt well to school despite their condition, parents may perceive greater challenges than the children experience. Our findings support using both parent-proxy and child reports in QOL assessments, as they show strong overall agreement but diverge in school functioning. While proxy reports reliably capture global QOL, self-reports remain essential for school-related evaluations. Future studies should examine ways to enhance parent-child communication about school experiences and explore cultural influences on reporting accuracy.

This study found significantly lower total QOL scores among Sabah's thalassaemia patients compared to Shafie *et al.* (2020) Malaysian cohort, but the results were consistent with recent data from India. (Chinnaiyan *et al.*, 2024). While our sample included patients aged 8 to 18 years, Shafie *et al.* (2020) included younger children (5 to 8 years). Age has been consistently identified as a significant predictor of quality of life (QOL) in paediatric thalassaemia research. Studies demonstrate that younger children typically report better QOL

scores, potentially due to developing disease awareness and emerging coping strategies (Varni *et al*., 2009). In contrast, adolescents frequently exhibit greater psychosocial distress as they navigate disease-related limitations on autonomy and social development (Treadwell *et al*., 2024). Our study's inclusion of age groups (8-18 years) appears to reflect this developmental trajectory, particularly in school functioning, where older participants' scores reflected accumulating academic and social challenges.

The present found that physical health scores were significantly higher than psychosocial health scores among transfusion-dependent thalassaemia patients. This pattern aligns with multinational research across Southeast Asia, including studies from Malaysia (Ismail *et al*., 2013; Shafie *et al*., 2020), Thailand (Sinlapamongkolkul & Surapolchai, 2020), and India (Chordiya *et al*., 2018). This persistent divergence suggests that while medical advancements have improved physical outcomes, psychosocial well-being remains suboptimal. The introduction of novel iron chelation therapies and optimised transfusion protocols has reduced physical complications, allowing patients to maintain near-normal activity levels. Despite physical health improvements, psychosocial care remains underprioritised, particularly in low-resource settings.

The school functioning scores were the lowest in psychosocial subscales, a trend also reported in other studies (Ismail *et al*., 2013; Kumcagiz & Cengiz, 2017; Shafie *et al*., 2020; Biswas *et al*., 2023; Permana *et al*., 2024). The low school functioning scores could be due to frequent blood transfusions, leading to missed school days or in severe cases, school dropout. School absenteeism may affect lesson retention, academic difficulties and increase stress levels among the thalassaemia children. In addition, there may be a lack of adequate resources in schools to support children with chronic illnesses and limited awareness among teachers and peers could lead to a lack of academic and social support. To address these challenges, out-of-hours treatment options could help reduce school absenteeism. Possible solutions include flexible nursing shifts to support evening or weekend outpatient clinics, mobile transfusion units or home-based transfusion services to minimise disruptions to school attendance, and collaboration between nurses and educators to train teachers on how to accommodate children with thalassaemia, creating a supportive learning environment to enhance both the academic and social well-being.

This study found a significant association between the physical health summary score and serum ferritin level. However, previous local studies identified different predictors for physical health summary score, including age, gender, absence of blood transfusion or chelation therapy, and occurrence of side effects from chelation treatment (Sazlina *et al*., 2015; Shafie *et al*., 2020). Unlike findings from other studies (Mufti *et al*., 2015; Khodashenas *et al*., 2021), this study found that patients with serum ferritin levels above 2500 ng/mL had better physical health scores. Thalassaemia patients with long-term iron overload often exhibit physical appearances such as darker skin, short stature and puberty delay. However, it has been reported that thalassaemia patients tend to accept their physical differences and cope with them over time (Mufti *et al*., 2015). This unexpected result may be attributed to patients' acceptance of their condition and their ability to cope and minimise differences, which may reduce the impact of physical differences on their perceived well-being. Additionally, it is possible that more frequent monitoring in patients with higher serum ferritin levels leads to better symptom management and an improved perceived quality of life. Despite adjusting for confounding factors, this unexpected finding may be influenced by other unmeasured variables, such as nutritional status and dietary intake, which were not included in this study. Future research should explore these factors in greater depth. Additionally, given that the number of patients with serum ferritin levels above 2500 ng/mL was small, further studies with a larger sample size are necessary to confirm these findings.

This study reveals that patients with divorced parents had significantly lower overall QOL scores and psychosocial scores than those from intact families. This extends beyond socioeconomic factors identified in recent research (Qiao *et al.*, 2024), highlighting family stability as a key determinant of well-being in thalassaemia care. However, the association between parents' marital status and QOL in thalassaemia patients has not been extensively studied. A previous study found that children with thalassaemia who had strong family support reported better QOL (Astarani *et al*., 2024). Divorce may negatively impact a child's overall well-being in several ways by causing emotional distress, as children may feel abandoned or experience family instability, causing disruptions in medical care due to uncoordinated schedules between separated parents and financial

strains, which may limit access to medical treatment and necessary care. Parental marital status may also reflect broader family dynamics, such as conflict or communication issues, which were not measured in this study. Future qualitative studies are needed to explore how changes in parental marital status affect thalassaemia patient's well-being in greater depth. As advocates, nurses play a crucial role in assessing patients' family histories and helping families navigate the challenges of long-term therapy. Establishing a helpline or online support group led by nurses could provide psychosocial support for thalassaemia patients and their families, helping to enhance their emotional well-being and resilience.

The type of iron chelation therapy was identified as another significant predictor for psychosocial health and total summary scores, consistent with findings from previous studies conducted in Malaysia (Shafie *et al*., 2020) and in India (Chordiya *et al.*, 2018). Patients who received combination therapy involving both oral and subcutaneous injection therapy had better psychosocial health and total summary scores. This benefit likely stems from more effective iron control, as combination regimens have been shown to maintain optimal serum ferritin levels (Alavi *et al.*, 2014), thereby reducing iron overload complications that impair physical and emotional well-being. While Silva and Peiris (2020) reported neutral QOL outcomes with combination therapy, this discrepancy may reflect differences in patient age distributions, treatment duration and psychosocial support system. Nurses play a critical role in chelation therapy by providing education, monitoring, administration and support. The implementation of nursing interventions such as structured education materials on indications, mechanisms, side effects and types of chelators; demonstrating proper administration techniques for oral or injectable chelators; develop personalized medication schedules based on the patient's lifestyle; and helping families apply for financial aid or insurance coverage for chelators may enhance patient's adherence to the chelation therapy. Mobile application for side effect tracking, medication reminders, and virtual peer support developed by a nurse may enhance accessibility and improve chelation therapy adherence.

Limitation

The study acknowledged several limitations that should be addressed. Firstly, this study did not examine important psychosocial factors such as illness acceptance, social support and family adjustment, which can influence patients' QOL. Including these variables in future research may provide a comprehensive understanding of the psychosocial aspects affecting thalassaemia patients' QOL. Secondly, illiterate patients were excluded from the study, which may introduce bias as their QOL experiences were not captured. A broader inclusion of patients, regardless of literacy level, would help ensure a more representative sample and a better reflection of the overall thalassaemia population. Lastly, since the study used a cross-sectional design, data was collected at a single time point, limiting its ability to assess long-term QOL outcomes. Despite these limitations, this study provides valuable insights into the QOL of paediatric transfusion-dependent thalassaemia patients in Sabah, a state with the highest thalassaemia prevalence in Malaysia. The findings serve as a baseline reference for future research, helping to shape more comprehensive studies and improve healthcare strategies for this population.

Recommendation

A nurse-led programme can be highly beneficial to address various aspects of patient care, including family counselling to support emotional well-being, financial assistance to ease the burden of treatment costs, peer support networks to enhance social integration, and guidance on school accommodations to ensure continued education. Integrating technology-based solutions such as digital platforms, virtual consultations, and online resources can improve accessibility and support for patients and families. Continuous assessment of patients' health outcomes, including QOL, can help nurses monitor patient well-being and make timely interventions to improve health outcomes. For future research, longitudinal design would provide more robust and insightful information regarding changes in patients' QOL over time. Research should include a wider range of psychological factors affecting QOL, including family adjustment, coping strategies, and social support systems. A mixed-method research design is recommended to capture detailed insights into the family dynamics and explore other psychosocial aspects that may not be easily quantified.

CONCLUSION

This study found that the QOL scores of paediatric transfusion-dependent thalassaemia patients in Sabah

were lower compared to previous studies conducted in Malaysia. Improvements in QOL, particularly in school functioning are necessary to enhance patient well-being. This study recommended considering QOL as an important factor in the management of transfusion-dependent thalassaemia patients, with a focus on reducing school absenteeism through the provision of out-of-hours thalassaemia treatment services. A nurse-led programme can be highly beneficial to address various aspects of patient care, including family counselling to support emotional well-being, financial assistance to ease the burden of treatment costs, peer support networks to enhance social integration, and guidance on school accommodations to ensure continued education. Integrating technology-based solutions such as digital platforms, virtual consultations, and online resources can improve accessibility and support for patients and families. Continuous assessment of patients' health outcomes, including QOL, can help nurses monitor patient well-being and make timely interventions to improve health outcomes.

Conflict of Interest

The authors declare that they have no competing interests.

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