Original Article

Predicting factors impact to quality of life of school age Thalassemic children in Indonesia

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ABSTRACT

Thalassemia is a major genetic blood disorder that considered a public health problem in Indonesia. While significant advances have been made in the clinical management of thalassemia children over the past few decades, our knowledge of the factors affecting the quality of life of thalassemic children is limited and some previous studies have shown contradictory results. The study aimed to analyze factors impacted to the QOL of school-age thalassemic children in Indonesia. A correlational analytic with a cross-sectional approach was conducted at a district hospital in Sumedang Indonesia from May to July 2017. The PedsQL generic core scale was used to assess 55 school-age thalassemic children with thalassemia major. Data were analyzed bivariate by using Pearson and Spearman Correlation Test and multivariate analysis used multiple linear regression to determine the factor that most impacting the QOL thalassemic children. The findings showed that the average of QOL of school-age thalassemic children was 66.54+12.85. There was a significant correlation between QOL with pre-transfusion Hb level (p=0,018, α=0,05), frequency of transfusion (p=0,000, α=0,05), nutrient status (p=0,000, α=0,05), family support (p=0,004, α=0,05), friends support (p=0,000, α=0,05) and adherence to iron chelation therapy (p=0,016 and α=0,05). Transfusion frequency is the most predicting factor that influences the QOL of school-age thalassemic children. It is suggested that effective transfusion scheduling, family and friends support, and providing adequate education for parents are essential in improving QOL thalassemic children in Indonesia.

Keywords: Factors, school-age children, thalassemia major, quality of life

Introduction

Thalassemia major is an inherited hematological single gene disorder [1] leading to anemia in affected children in the world that represents a major public concern in Southeast Asia, including Indonesia [2, 3]. Regular blood transfusion and the use of iron-chelating agents have been the mainstay of supportive treatment in thalassemia [4, 5]. Indonesia is one of the countries in the thalassemia belt. In Indonesia thalassemia mayor, is more prevalent and as a serious health problem; approximately 3-10% of people are carrying the genes of thalassemia due to abnormal production of hemoglobin [6]. The thalassemia foundation in Indonesia mentioned that the number of thalassemia patients in 2015 was 7,029 spread across 23 provinces [7], and there were 3,300 people with thalassemia in West Java alone in January 2016 [8].

Thalassemia major causes a lot of problems for the thalassemic children due to the defect of various organs because of the illness itself or the treatment being given [9-12]. Children with thalassemia major who do not get regular blood transfusions can experience severe physical anemia, the effects of which include Cooley's faces characterized by protrusion of the forehead, depression of the nasal bone, mongoloid-like eye tendencies, maxillary hypertrophy, maxillary malocclusion, and enlarged spleen [10]. Besides, the child will experience growth retardation and skeletal changes due to bone marrow expansion, resulting in the risk of bone disability of the foot. Hemosiderosis or accumulation of

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iron also causes various complications such as cardiac, endocrine, and hepatic complications [2, 10]. Physical changes such as bone deformities and short stature cause impaired self-image, low self-esteem, anxiety, feel socially isolated with conditions of illness, may also be depressed, and other possible effects [12, 13]. Psychological and social impacts will greatly affect school-age thalassemic children in the form of socialization disorders and disorders with social groups [9, 12].

These problems overall impair the QOL of thalassemic children [9, 12, 14]. A study conducted in Sumedang District hospital Indonesia which involved 46 respondents (school-age thalassemia children), found that most of the children had bad QOL (67.4%) [15]. The QOL of school-aged thalassemic children with thalassemia major in the hospital may be influenced by many factors, however, there has been no research related to this in our study setting. Factors associated to QOL of thalassemic children that need to be investigated in this current study are family support, pre-transfusion Hb rate, frequency of transfusion and the nutritional status of children, along with factors that have not been studied, including the support of friends and adherence to undertaking iron chelation therapy. Therefore, this study needs to analyze factors that relate to the QOL of school-age children with thalassemia major at the Thalassemia Clinic of the Sumedang District Hospital following the characteristics and conditions of each thalassemic child. A good understanding of predicting factors that affect QOL among thalassemic children may have an impact on the development strategies and programs such as genetic counseling, social support programs to improve treatment outcomes and the QOL of the thalassemic children. Given the limited studies in this area in Indonesia, these study aims were to identify factors impacted QOL among school-age thalassemic children with a focus on parents' perspective.

Material and Methods

A descriptive, correlational design was employed. The study was conducted in the Thalassemia Clinic Sumedang District Hospital Indonesia from May to July 2017. The numbers of school-age thalassemia children were 55 people. The inclusion criteria for this study were children in the age group of 8-12, diagnosed with thalassemia major, registered and following up at the hospitals at least past six months. The total sampling was used to select a sample of 55 school-age thalassemic children. All school-age thalassemic children and their parents agreed to participate in the study.

The Pediatric Quality of Life Inventory (Ped-QOL) 4.0 Generic Core Scale (Indonesian version) for 8-12 years old children was used for measuring the QOL of school-age thalassemic children. The Indonesian version of this questionnaire had already been validated[16]. The Internal consistency reliability of the Ped-QOL 4.0 Generic Core Scale approached 0.90 for self-report. The Ped-QOL 4.0 encompasses the essential core domain for QOL measurement: Physical functioning (8items); Emotional functioning (5items); Social functioning (5items) and School functioning (5items). The factors studied related to QOL of school-age thalassemia children were pre-transfusion Hb levels, frequency of transfusions, nutritional status, family support, peer group support, and adherence to iron chelation therapy.

Written parental informed consent and the thalassemic children's assent were gained before participating in the study. At the beginning of collecting data, all respondents were informed of the aims of the study and were assured that all responses would remain confidential. To assess the QOL of thalassemic children, the Ped-QOL questionnaires were to be completed independently by school-age children (8-12) years.

Data were analyzed by Microsoft Excel 2010 and SPSS Program version 13.0. General characteristics of the thalassemic children were presented in terms of percentage, mean, and standard deviation. For QOL, both total QOL score and summary scores were presented in terms of mean and standard deviation. Bivariate data analysis used Pearson's and Spearman's correlation tests, and multivariate analysis used multiple linear regressions. Linear regression analysis was used to identify independent predictors of high QOL scores among all studied factors. P-value was considered significant if P was 0.05.

Ethical clearance was gained before the start of this study from the Health Research Ethics Committee Faculty of Medicine Universitas Padjadjaran Bandung. The ethical approval number is 800/UN6. C.10/PN/2017. Confidentiality was warranted by restricting access to the names of respondents.

Results

The characteristics of the respondents in this study are illustrated in Table 1. Parents of these children were asked to fill the PedQL questioners.

Table 1: Demographic Characteristics school-age thalassemic children (n = 55)

Variable	f	%
Sex		
Male	31	56.4
Female	24	43.6
Child education		
No school	11	20.0
Primary school	44	80.0
Middle school	-	-
High School	-	-
College	-	-
Father Education		
No school	-	
Primary school	17	30.9
Middle school	12	21.8
High School	19	34.5
College	7	12.7
Mother Education		
No school	-	
Primary school	16	29.1
Middle school	17	30.9

High School	17	30.9
College	5	9.1

It can be seen from table 1 of the respondents, 56.4% are male and 80% were educated at elementary school level (SD). Most fathers of the respondents (34.5%) had a high school education, while 30.9% of mothers of the respondents had the same education level. The cumulative average scores of QOL based on domains are illustrated in table 2

Table 2. The mean PedQOL Score of school-aged thalassemic children (n = 55).

Domain	Mean ± SD	Min	Max
Physical Functions	66.48 ± 16.38	12.50	90.63
Emotional Functions	72.00 ± 14.42	30.00	100.0
Social Functions	78.18 ± 18.49	20.00	100.0
School Functions	49.55 ± 14.31	20.00	75.00
The total Score	66.54 ± 12.85	21.74	89.13

As can be seen from Table 2, the total QOL score was found to be (66.54±12.85) with the highest in the social (78.18±18.49) and emotional (72.00±14.42) domains followed by physical (66.48±16.38) and lastly the lowest score in school domain (49.55±14.31). To identify the relationship of each independent variable with the dependent variable, bivariate analysis was conducted by using Pearson's Test and Spearman's Test. It can be seen in Table 3.

Table 3. Results of the bivariate analysis on variables related to QOL school-age thalassemic children

No.	Independent Variable	Dependent Variable	
		p	*r*
1	Pretransfusion Hb levels	0.018	0.318
2	Transfusion frequency	0.000	0.571
3	Nutrition status	0.000	0.465
4	Family support	0.040	0.278
5	Friend support	0.000	0.556
6	Adherence to iron chelation therapy	0.016	0.323

Based on Table 3, pre-transfusion Hb levels, frequency of transfusion, nutritional status, family support, friend support, and adherence to taking medication for iron chelation therapy are significantly correlated with the quality of life of the school-age sufferers of thalassemia major. Meanwhile, the results of multiple linear regression analyses are presented in table 4 below.

Table 4 Summary of multiple linear regression analysis results

Free Variable	Regression Coefficient	tcount	Sig.	Colinearity	
				Tolerance	VIF
Constant	28.511				
Transfusion frequency	10.321	3.573	0.001	0.836	1.196
Friend support	0.546	3.476	0.001	0.836	1.196
F count	= 20.872	Sig. 0.000			
R^2	= 0.445				
Adjusted R^2	= 0.424				

Table 4 shows that the regression coefficient of the transfusion frequency variable (10.321) is greater than that of the variable of friend support (0.546). It is concluded that the frequency of transfusion is the most influential factor in QOL of thalassemic children.

Discussion

Assessment of QOL among school-age thalassemic children indicated that the average score of QOL was 66.54. The average score of QOL in the domain of school functions occupies the lowest position comparing other domains. This low score could be related to frequent visits to the hospital for blood transfusions or treatment of iron chelation therapy, so that children cannot attend school because they feel weak and do not have enough energy to follow the lessons at school. This is supported by the children's expressions that if they are sick then they cannot go to school, making the frequency of not going to school high enough. Children who do not attend school have decreased cognitive abilities that adversely affect their quality of life. These study results are in line with earlier studies [12, 16]. Meanwhile, psychosocial functions had a higher score than physical and school function which differ from results of previous studies [17, 18]

The results indicated that there was a positive correlation between the frequency of transfusion and the children's quality of life. Regular transfusion has a very meaningful impact on the children, as they can assume normal life up to 10 or 12 years old or the age of puberty [2]. This current study is in line with an earlier study [19]. However, other previous studies [12, 20, 21] found that there was no relationship between the frequency of transfusions and the QOL of thalassemic children.

Another significant finding of this study showed that there was a relationship between family support and the QOL of thalassemic children. This result is in line with an earlier study conducted in Indonesia [21], leading to the conclusion that the greater the family support, the greater the quality of life of the children. Parents need to learn additional tasks related to caring for children with thalassemia, which can affect the parents' psychology [19, 22, 23]. Nurses as family facilitators play an active role in motivating families to improve their family ability independently.

This current study also found that there was a positive correlation between adherence to take iron chelation drugs with the QOL of children with thalassemia major. Similarly, a previous study [12] indicated that giving iron chelation drugs and side effects from iron chelation treatment were predictors of the QOL of thalassemic children. [24]. Nurses need to enhance thalassemic children's adherence to drink iron chelation drugs by educating thalassemic children and their families about iron chelation drugs and increase family support.

The results of this study identified that the most influential factor to the QOL of thalassemic children was the frequency of transfusion. Blood transfusion is the main therapy for children with thalassemia major [12]. Therefore, the nurse needs to educate the family about the importance of timeliness of the child's schedule for transfusion and the importance of maintaining the condition of the child. The nurse also needs to coordinate and organize health services with other health teams such as Community Health Insurance to facilitate blood transfusion according to the needs of the patients.

Conclusion and recommendations

Thalassemia major and its treatment have a negative impact on school-age thalassemic children' QOL, especially in the school functioning domain. The findings of this study highlight the blood transfusion frequency has the most significant negative impact on QOL. In light of this, and considering the prevalence of thalassemia in Indonesia, our study suggested that it is essential to develop a model of integrated thalassemia care in Indonesia that involved health care providers and thalassemic children's parents.

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Competing Interest:

The authors declare that there is no conflict of interest.

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