



Original Research

## Quality of Life of Preschool-Age Children with Thalassemia Major

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### ABSTRACT

**Introduction:** Health problems caused by thalassemia disease process and treatment affect patients who suffer from this disease, including pediatric patients who are in their preschool age. These problems can interfere with the child's physical, emotional, social, and school functions. This study aimed to describe the quality of life of preschool-age children with thalassemia major in Sumedang and Garut areas.

**Methods:** In this study, a quantitative descriptive design was applied to a sample of 63 parents of children with 3-6 years old children with thalassemia who were recruited using the total sampling approach. The quality of life as a variable was measured using the PedsQLTM 4.0 Generic Core Scales for parent-proxy reports. Data collected were then analyzed for the mean score and categorized into good or poor quality of life, which were then expressed in frequencies and percentages.

**Results:** Results showed that the mean score of quality of life was 75.94, with the mean scores for physical function, emotional function, social function, and school function of 75.79, 71.98, 87.46, and 60.95, respectively. The majority of preschool-age children with thalassemia have poor quality of life. The dimension that receives the most negative influence is the school function.

**Conclusion:** It is suggested that nurses can improve the quality of life of these children by identifying the impaired functions and providing the appropriate care, such as facilitating the needs of these children to receive blood transfusions in hospitals.

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## 1. INTRODUCTION

Thalassemia is one of the genetic diseases that become major concerns in various parts of the world, including in Mediterranean, Middle East, and Asian countries (World Health Organization (WHO), 2006). Approximately 5% of the world population are carriers of thalassemia and about 1.1% of couples around the world are at risk of having a child with thalassemia (Abu-Shaheen et al., 2020). In Indonesia, the prevalence of thalassemia carriers is around 3-8%. If it is assumed that the percentage of thalassemia is 5%, the birth rate of infants with thalassemia would be 23 per 1,000. Thus, with the population size of 240 million, it is estimated that 3,000 babies are born with thalassemia in Indonesia each year (MOH, 2012). Based on the data from the Indonesian Thalassemia Foundation (Yayasan Thalasemia Indonesia [YTI], 2017), the number of thalassemia cases is increasing from year to year. West Java is the province with the highest number of thalassemia cases in Indonesia, and Sumedang and Garut are among the top 10 cities/districts with the highest number of thalassemia patients (YTI, 2017). As a result, the quality of life of preschool-aged children with thalassemia major in the Sumedang and Garut areas must be described.

Thalassemia is a genetic disease in the form of red blood cell disorders that is characterized by the destruction or loss of one of the globin chains that normally make up normal hemoglobin. The most common type of thalassemia found in children is thalassemia major that requires lifetime blood transfusions. This disease can be diagnosed from an early age through the presence of symptomatic anemia at the age of 6-12 months (Jafari-Shakib et al., 2016).

Thalassemia in children can lead to various health problems that will affect their life. These health problems will lead to the disruption of the physical, emotional, and social functions of the child, and will also disrupt his or her school activities. Children with thalassemia may have similar health problems. However, the impacts may be differ depending on the stage of development of the child. One of the most important developmental stages in a child's

life is the preschool age. This stage is characterized by the peak of development and any chronic pain experienced by the child during this period will have a big impact on their life, especially in his or her development and social development (Potter & Perry, 2014).

The negative impact that arises due to thalassemia will affect the quality of life of the affected children and lead to a low quality of life. Research conducted in Egypt discovered that there is a decrease in quality of life in all functions among children with thalassemia (Ismail et al., 2018). Other studies also report that the quality of life of thalassemia children in Aceh is low (Nikmah, 2018).

Assessing a child's quality of life can become a way to identify the impact of a disease and its treatment (Sharma et al., 2017). Pediatric Quality of Life Inventory TM 4.0 Generic Core Scales, or commonly referred to as the PedsQL, is one of the standard instruments used for measuring children's quality of life. This scale assesses children's quality of life from four dimensions: physical function, emotional function, social function, and school function (Varni, 2019). The physical function looks at the impact of physical changes in the form of its influence on the child's physical activities. The emotional function assesses the feelings or emotions that the child feels about the disease and its treatment, while the social function looks at the social activities and development of affected children in terms of their interactions with their peers and their environment. The school function assessed how the illness interfere the child's school activities in affected children who are already in school and, in the preschool-age children, how the playgroup activities are affected by the disease.

Results from a preliminary study in hospitals Sumedang and Garut on parents of children with thalassemia aged 4-6 years demonstrated several issues in these children, such as slower growth; restlessness that is sometimes experienced by the child during transfusion especially when the blood given is not really suitable; and, for those who are already in school, absence from school in order to receive transfusion in the hospital. These indicate disruptions in the functions in the dimensions of the quality of life.

Understanding the quality of life is important to evaluate the effectiveness of health services and to determine the benefits of the measures provided to patients (Mikael & Al-Allawi, 2018). In order to provide the best care a nurse needs to understand the quality of life of thalassemia children in order to provide appropriate interventions according to the child's needs. However, there are not many studies that specifically discuss the quality of life of thalassemia children, especially those that involved preschool-age children with thalassemia as the subjects. Therefore, this study aimed to describe the quality of life of preschool-age children with thalassemia major in Sumedang and Garut areas.

## 2. METHOD

### 2.1 Design

This was a descriptive quantitative study with the quality of life of preschool-age children with thalassemia major as the variable.

### 2.2 Population

#### 2.2 Population, Samples, and Sampling

The population in this study was parents of children with thalassemia major aged 3-6 years old in Sumedang and Garut areas. Total sampling was used to collect the sample of 63 parents, consisting of 22 parents from Sumedang and 41 parents from Garut

### 2.3 Instruments

Data were collected using questionnaires that include a questionnaire on respondent data, PedsQL 4.0 Generic Core Scales for parent-proxy reports for groups of children aged 2-4 years and 5-7 years. Meanwhile, the PedsQL questionnaire used consisted of 23 questions that were divided into four multidimensional scales to measure physical function (8 items), emotional function (5 items), social function (5 items), and school function (5 items). As for the parent-proxy report for groups of children aged 2-4 years in school function, there were only three items in the questionnaire.

### 2.4 Procedure

Data collected through the respondent data questionnaire included demographic

data and clinical characteristics that include gender, parents' education, parents' occupation, age at diagnosis of thalassemia, age at first transfusion, and frequency of blood transfusion.

### 2.5 Analysis

Data obtained were then analyzed using the mean score of total scores, through the addition of the scores, which were then divided by the number of question items. The scoring for each question item was reversed to a value of 0-100 using the following Likert scale: Scale 0 (never) gained a score of 100; 1 (almost never) gained a score of 75; 2 (sometimes) gained a score of 50; 3 (often) gained a score of 25; and 4 (almost always) gained the score of 0. Quality of life was categorized as good or poor based on the mean score obtained from the total score calculation of the child's quality of life

### 2.6 Ethical Clearance

This research was previously approved by the Health Research Ethics Commission of Padjadjaran University, with the letter number: 356/UN6.KEP/EC/2019...

## 3. RESULT

Results of the analysis are presented in two tables. The first table (Table 1) lists the characteristics of the respondents while the second table (Table 2) presents the mean score of the quality of life.

Table 1 on the characteristics of the preschool-age children with thalassemia major preschoolers in Sumedang and Garut areas shows that there were more boys (n=37, 58.7%) compared to girls (n=26, 41.3%). The majority of fathers of these children were self-employed, which comprised 61.9% (n=39), with most graduated from junior high school or equivalent (n=24, 38.1%). Meanwhile, the majority of the mothers were housewives (n=52, 82.5%) with most of them graduated from junior high school or equivalent (n=30, 47.6%). It was also demonstrated that the age when the child was diagnosed with thalassemia and the age at first transfusion of the majority of children were mostly in the range of 0-1 years (n=59, 93.7%). Most children had transfusions once a month (n=32, 50.8%) with the majority had an Hb level of <9 g/dl prior to transfusion (n=55, 87.3%).

Table 2 presents the mean quality of life (QoL) of the respondents. In this study, the quality of life dimension with the highest score was the social function with a mean score of 87.46 while the dimension

with the lowest score was the school function with a mean score of 60.95. Overall, the mean score of the respondents' quality of life was 75.94.

Table 1. Respondent Characteristics (n=63)

<b>Chararacteristics</b>	<b>N</b>	<b>%</b>
Gender		
Boy	37	58.7
Girl	26	41.3
Faher's Education		
Elementary/equivalent	14	22.2
Junior High School/equivalent	24	38.1
Senior High School/equivalent	20	31.7
University	5	7.9
Father's Occupation		
Civil Servant	1	1.6
Self-Employed	39	61.9
Unemployed	0	0
Others	23	36.5
Mother's Education		
Elementary/equivalent	15	23.8
Junior High School/equivalent	30	47.6
Senior High School/equivalent	13	20.6
University	5	7.9
Mother's Job		
Civil Servant	1	1.6
Self-Employed	4	6.3
Unemployed/Housewife	52	82.5
Others	6	9.5
Age at Diagnosis of Thalassemia		
0-1 years old	59	93.7
2-4 years old	3	4.8
> 4 years old	1	1.6
Age of First Transfusion		
0-1 years old	59	93.7
2-4 years old	2	3.2
> 4 years old	2	3.2
Transfusion Frequency		
Once a week	0	0
Once every two weeks	9	14.3
Once every three weeks	22	34.9
Once a month	32	50.8
Pre- transfusion Hb level		
< 9 g/dl	55	87.3
9 g/dl	8	12.7

Table. 2 Average R score of Quality of Life (n=63)

<b>QoL Dimension</b>	<b>Average</b>
Physical Function	75.79
Emotion Function	71.98
Social Function	87.46
School Function	60.95

#### 4. DISCUSSION

This study reveals that the mean total score of quality of life in preschool-age with thalassemia is 75.94. From the perspective of the mean score of the quality of life, the quality of life is considered poor when the score is less than 80 (Nikmah, 2018). The school functions score of 60.95, which indicated that children's ability to focus on completing schoolwork was not in line with children their age, represented the lowest quality of life in this study. Then, the scores for physical ability and emotional function were 71.80 and 75.79, respectively, with a mean value below 80. As a result of these findings, it can be inferred that the child requires a lot of assistance in carrying out his daily activities and have some emotional problem to express his fear, sadness, or anger.

Therefore, it can be said that the quality of life of the preschool-aged thalassemia patients in this study is lower than the quality of life of generally healthy or normal children. A previous study also presents a similar result, with children with thalassemia having a lower QoL than normal children (Mariani et al., 2014). The mean total quality of life score in this study is lower than the score obtained in a previous study in India (Dhirar et al., 2016; Sharma et al., 2017). However, the quality of life in this study is higher than those reflected in studies conducted in Egypt and other parts of Indonesia (Adam et al., 2017; Mariani et al., 2014; Nikmah, 2018).

The low quality of life can be influenced by many factors, such as age of onset of anemia of before 2 years old, age of first transfusion of before 4 years, and a pre-transfusion Hb level of <9 g/dl (Mikael & Al-Allawi, 2018). This is in line with the findings in this study, showing that most respondents were diagnosed with thalassemia at the age of 0-1 years, with the age of first blood transfusion of 0-1 years and a pre-transfusion Hb level of <9 g/dl.

With reference to Table 2, it can be observed that among all dimensions of quality of life, the mean score of the school function is the lowest (60.95). School function can be disrupted due to child's absence from school and the decline in the

academic achievement linked to the child's need for routine transfusions (Ismail et al., 2018). A previous study conducted by Yasmeen & Hasnain, (2018) showed the same low school function among children with thalassemia in Pakistan. This may relate to the lack of energy that the child has to go to school, which added the percentage of absence from school and reduced quality of life. Although transfusion can be provided after school hours, it is still difficult to avoid being absent from school when the child lives in a rural area that does not yet have nearby transfusion facilities or in places where transfusion service is not provided at the nearest health care facility. In preschool children, the school function refers to non-formal education activities, such as kindergarten because children in these age are not included into the group of children that have to follow compulsory education.

The emotion function is the dimension with the second lowest score of 71.98. Emotional disturbances in preschool-age children with thalassemia link to the child's response to the illness. Poor quality of life in the emotional function may be influenced by the length of time suffering from thalassemia, which is less than 10 years, and the characteristics of preschool-age children (Sazlina et al., 2015). Table 1 demonstrates that the majority of children with thalassemia (93.7%) were diagnosed when they were 0-1 years old. Younger patients have little experience and do not understand the disease they have to live with and this can affect their emotional state. The preschool-age children usually experience fear towards anything that they perceive will harm them and they may act more aggressively due to this fear. In older children, the past experience with the disease and support from their parents, family, and peers may ease up this fear. In addition, older children also have sufficient knowledge about thalassemia, which can affect their quality of life (Hamdy et al., 2021).

This study also discovered that the physical function, with the mean score of 75.79, is also poor among preschool-age children with thalassemia. The poor quality of life in physical function can be influenced by several factors, such as the clinical characteristic aspects. A previous study also reported that physical function is associated with thalassemia patients who receive regular

blood transfusions in the last three months (Thavorncharoensap et al., 2010). This supports the results of this study, that the poor quality of life in physical function may be due to the fact that most respondents receive routine transfusion once a month (50.8%).

The dimension with the highest score for quality of life is the social function with a mean score of 87.46. This result is in line with a study conducted in Aceh (Nikmah, 2018). Preschool-age children with thalassemia major do not experience disturbances in their social functions because they can still socialize with their peers even when they are sick.

## 5. CONCLUSION

Preschool-age children with thalassemia major (3-6 years) in Sumedang and Garut areas have poor quality of life in all dimensions. The functions that are most affected by the disease are, in ascending order, social function, physical function, emotional function, and school function. School function is the most affected dimension of quality of life because of the absenteeism of children with thalassemia due to the need for treatment in hospital or being unwell. The social function is the least affected dimension as these children are still able to socialize and interact with their peers. Thus, it is suggested that to improve the quality of life of preschool-age children with thalassemia, nurses should be able to identify the impaired functions and provide appropriate care to these children, such as through facilitating the needs of the child when he or she undergoes blood transfusion in the hospital.

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## 7. CONFLICT OF INTEREST

The Authors declare that there is no conflict of interest.

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