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Quality Of Life of Children with Thalassemia in Indonesia: Review

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Abstract

Background: *Thalassemia is an inherited disorder of hemoglobin (Hb) synthesis. Treatment of thalassemia has not yet reached the level of cure. Globally, the management of thalassemia is symptomatic in the form of lifelong blood transfusions. Data on children with thalassemia major in Indonesia has not been widely published. However, various studies clearly showed growth and development disorders and behavior that greatly affected the quality of life of children with thalassemia. Blood transfusions and lifelong use of drugs often lead to feelings of boredom of treatment, not to mention physical changes and the feeling of being different from relatives or friends that will cause the feeling of inferiority. They often drop out of school and do not find work, which causes highly severe psychosocial effects.*

Objective: *This paper aims to provide an overview of the quality of life of children with thalassemia in Indonesia so that it can be used as a reference in providing appropriate management regarding the quality of life of children with thalassemia.*

Methods: *This literature search used google scholar and pubmed for complete publications in 2010-2020 with the keywords "quality of life", "thalassemia children," and "Indonesia".*

Result: *There were six research articles related to this search. Indonesia's average quality of life for children with thalassemia ranged from 50% to 67.2%.*

Conclusion: *Of the four domains assessed, school function had the lowest average, followed by emotional, physical and social function. Child health services need to optimize children's functional abilities and quality of life.*

Keywords*: Children; Indonesia; Thalassemia; Quality of Life*

INTRODUCTION

Thalassemia is an inherited hemoglobin disorder (Hb) synthesis, particularly globin chains (Bains, 2020; Bakthavatchalam, 2019; Batool et al., 2022). This genetic disease has various types and frequencies in the world. The clinical manifestations vary from asymptomatic to severe symptoms. Data from the World Bank shows that 7% of the world's population are carriers of thalassemia traits. Every year about 300,000-500,000 newborns are

accompanied by severe hemoglobin abnormalities, and 50,000 to 100,000 children die from thalassemia β; 80% of this amount comes from developing countries (The Act of Health Minister on Thalassemia Management, 2018; Sharma et al., 2017; Torcharus & Pankaew, 2011)

Indonesia is one of the countries in the world's thalassemia belt, namely a country with a high frequency of thalassemia genes (number of carriers). It is evident from epidemiological studies in Indonesia, which found that the frequency of the beta-thalassemia gene ranges from 3-10%.

Data from all teaching hospitals only registered around 7670 thalassemia major patients throughout Indonesia. This figure is still much lower than the estimated actual number of data from the Center for Thalassemia, Department of Child Health, FKUI-RSCM; until May 2014, there were 1,723 Thalassemia patients with the largest age range between 11-14 years. The number of new patients continues to increase to 75-100 people/year, while the oldest age of patients to date is 43 years (The Act of Health Minister on Thalassemia Management, 2018).

Treatment of thalassemia has not yet reached the level of cure. Worldwide, the management of thalassemia is symptomatic in the form of lifelong blood transfusions (Shafie et al., 2020; Torcharus & Pankaew, 2011). The need for 1 child with thalassemia major with a body weight of 20 kg for blood transfusion and adequate iron chelation costs around Rp. 300 million per year. This amount does not include the costs of laboratory examinations, monitoring, and management of complications that arise (The Act of Health Minister on Thalassemia Management, 2018).

Data regarding the condition of children with thalassemia major in Indonesia has not been widely published, but various studies clearly showed the growth and development of disorders and behavior which are later related to the high incidence of depression, anxiety and other psychosocial disorders which greatly affect the quality of life of children with thalassemia (Ankush et al., 2018; Kaheni et al., 2013). In Indonesia, a study in 2009 on adolescents aged 13-18 with thalassemia major found that 50.5% of adolescents have a poor quality of life (The Act of Health Minister on Thalassemia Management, 2018).

Children with thalassemia major are at risk of experiencing delays in cognitive development, impaired communication, motor, adaptive, or socialization than normal children. In addition, there may also be growth problems or disorders such as short stature, late puberty, and behavioral and emotional problems (Thiyagarajan et al., 2019).

Developmental disorders can range from mild to severe, temporary to permanent, resulting from the incurable medical condition of thalassemia; repeated transfusions, which are tiring and traumatic, complications of the disease, and limitations to daily activities at school, play area, or workplace (Yasmeen & Hasnain, 2018).

Blood transfusions and lifelong use of drugs often lead to feelings of boredom of treatment, not to mention physical changes and the feeling of being different from relatives or friends that cause feelings of inferiority. They often drop out of school and do not find work, which causes highly severe psychosocial effects (Hassan & Azzab, 2016).

Child health services need to optimize children's functional abilities and quality of life so that children with thalassemia can grow into productive adults. Pediatric Quality of Life InventoryTM (PedsQLTM) can be used to assess a child's quality of life over time (Arian et al., 2020).

Based on the background above, this paper aims to provide an overview of the quality of life of children with thalassemia in Indonesia so that it can be used as a reference in providing appropriate management regarding the quality of life of children with thalassemia.

METHOD

This literature search used google scholar and Pubmed for complete publications in 2010-2020 with the keywords "quality of life", "thalassemia children," and "Indonesia". The inclusion criteria were the measurement of quality of life using the Pediatric instrument Quality of Life Inventory TM (PedsQLTM) in children aged 2-18 years suffering from thalassemia in various regions in Indonesia.

There were 6 research articles related to this search. The articles were reviewed by looking for similarities and differences, providing views, comparing and summarizing, and drawing conclusions. The details can be visualized in the Figure 1 below:

(see figure 1)

RESULT

(see table 1)

DISCUSSION

Physical functions included in the quality of life assessment illustrate the child's ability to carry out daily activities independently. The emotional function assessment describes the child's ability to express anger, sadness, and fear. Social function assessment describes a child's ability to interact with peers and get along with friends at school. Assessment of school functions describes a child's ability to do the tasks given at school (Kavitha & Padmaja, 2019; Mardhiyah et al., 2020; Nikmah & Mauliza, 2018).

The result of the 6 studies above showed that the social function of children had the highest average of the other three functions while the school function had the lowest average of the other three functions.

School function is the lowest domain in assessing the quality of life. It is due to obstruction of school activities and decreased academic achievement scores due to the treatment of thalassemia (Isworo et al., 2012; Mariani et al., 2014; Nikmah & Mauliza, 2018; Wahyuni et al., 2011). This education problem needs to be solved with more concern for children with chronic medical conditions. Children are expected to continue to attend regular school education according to their abilities. The school is also informed about the patient's medical condition, the need for regular school permission for transfusions, and the child's susceptibility to disease, and the school is expected to support the treatment of these patients.

An emotional function is the second function after school function. Disturbance of the emotional domain is probably caused by the thalassemia sufferer's physical condition, which causes its burden (a stress condition). It can affect immunological psychoneurotics. Thus, if the physical condition is considered stress, a substance resembling beta-carboline, a GABA antagonist, causes a decrease in the number (down-regulate) of GABA receptors. It then causes reduced resistance to anxiety and eases reactions to stress (Isworo et al., 2012; Kavitha & Padmaja, 2019; Mardhiyah et al., 2020). Emotional dysfunction is influenced by various things, such as feeling depressed during diagnosis, therapy that must be undertaken regularly every month and skipping school because of having to undergo therapy (Nikmah & Mauliza, 2018). Sources of stress and fear for school-age children can come from the school environment, where experiences that cause stress include ranking competition with classmates, being recognized by teachers, labeling, being unable to learn, and worrying about not passing exams which can cause emotional discomfort. Most of the fear of school-age children is related to school and family (Hastuti, 2014). Psychosocial support from families is expected to reduce emotional problems in patients with beta-thalassemia major. Further, it is explained that psychosocial support reduces emotional distress, increases the effectiveness of iron chelation and strengthens coping strategies to improve everyday life (Armina & Pebriyanti, 2021; Halim-Fikri et al., 2022; Pranajaya & Nurchairina, 2016; Ramadhanti et al., 2020).

Physical function is the third function that is impaired after school function. The physical changes of the subject were very striking due to chronic anemia and iron deposition in the organs. Physical changes occur in the form of facial bone deformity, splenomegaly, bone marrow expansion, short stature, and various symptoms caused by hemolysis. Different appearance is an important factor that affects personality development, such as a lack of self-image, embarrassment, and refusal to socialize and go to school (Kamil et al., 2020; Nikmah & Mauliza, 2018).

The social function had the highest average score between 60 - 80% among physical, emotional and school functions. It illustrated that the social function of children suffering from thalassemia was still quite good compared to the other three functions. Children could still socialize and adapt to peers or friends at school with all their limitations, such as the physical changes resulting from

treatment and the limitations of daily activities. Limited physical activity makes the subject unable to do things that can be done by healthy normal peers (Halim-Fikri et al., 2022; Nikmah & Mauliza, 2018).

CONCLUSION

Based on the result of this study, it can be concluded that the mean quality of life of children with thalassemia in Indonesia ranged from 50% to 67.2%. School function had the lowest mean of the four domains assessed, followed by emotional, physical, and social. Health services that could improve the quality of life of thalassemia patients could be provided optimally with effective and efficient management strategies and involve all sectors, both formal and informal so that the quality of life for children with thalassemia would not be different from that of the normal children.

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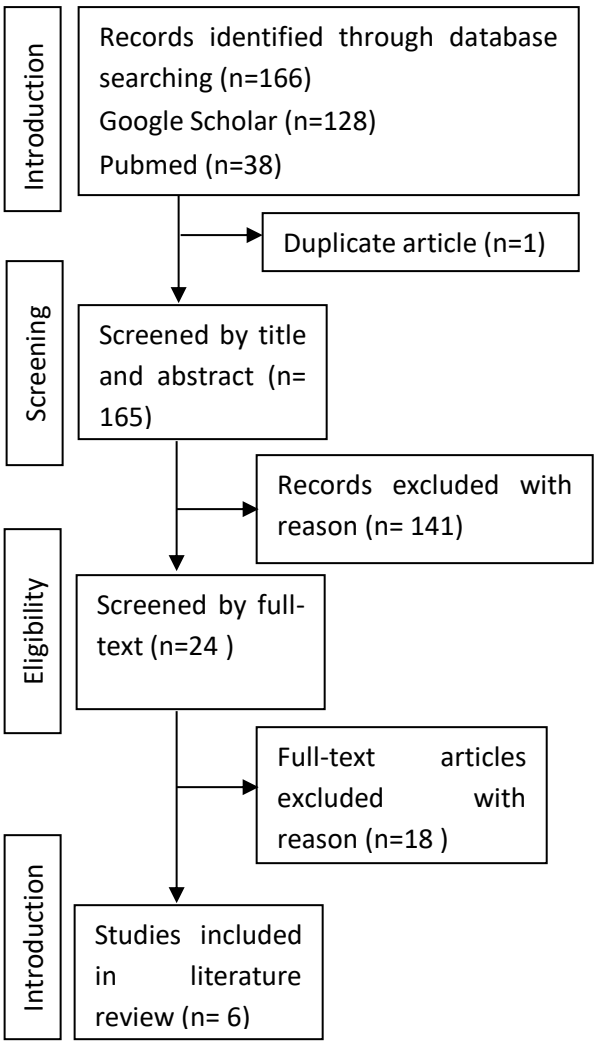

Figure 1. Prisma flow diagram

Table 1. Research in various regions in Indonesia regarding the quality of life of children with thalassemia

No	Title (years)	Method	Result	Conclusion
1	Quality of life assessment of children with thalassemia (2011). Wahyuni et al.	Children aged 5-18 years who suffer from thalassemia at Adam Malik Hospital, Medan	The Mean of physical function: 53.1% The Mean of emotional function: 50.9% The mean of social function: 62.5% The mean of school function: 36.2% The mean quality of life: 50.9%	The highest mean is a social function, and the lowest mean is a school function
2	Hemoglobin levels, nutritional status, food consumption patterns and quality of life of thalassemia patients (2012). Isworo et al	32 children with thalassemia aged 6-15 years in Banyumas Regional Hospital	The mean of physical function: 64.9% The mean of emotional function: 63.9% The mean of social function: 81.4% The mean of school function: 60% The mean of quality life: 67.2%	The highest mean is a social function, and the lowest mean is a school function
3	The effect of the thalassemia education package on the quality of life of thalassemia children (2014). Hastuti.	14 children aged 8-18 years at Abdul Moeloek Hospital Bandar Lampung	The mean of physical function: 66% The mean of emotional function: 61.4% The mean of social function: 79.29% The mean of school function: 53.9% The mean quality of life: 65.36%	The highest mean is a social function, and the lowest mean is a school function
4	Analysis of factors that affect the quality of life of children with beta-thalassemia major (2014). Mariani et al	84 children aged 5-18 years at RSU Kota Tasikmalaya and Ciamis	The mean of physical function: 60.86% The mean of emotional function: 57.61% The mean of social function: 61.46% The mean of school function: 54.52% The mean quality of life: 50.9%	The highest mean is a social function, and the lowest mean is a school function
5	Factors related to the quality of life of thalassemia children (2016). Pranajaya and Nurchairina.	102 children aged 5-18 years at Abdul Moeloek Hospital Bandar Lampung	The Mean of physical function: 65.72% The mean of emotional function: 61.72% The mean of social function: 70.34% The mean of school function: 56.01% The mean quality of life: 62.75%	The highest mean is a social function, and the lowest mean is a school function
6	Quality of Life for Thalassemia Patients based on the *Pediatric Quality of Life Inventory* 4.0 *Generic Core Scales* in the Nursing Room of Cut Meutia Hospital, North Aceh (2018) Nikmah and Mauliza.	41 children aged 2-18 years in the pediatric ward of Cut Meutia Hospital, Aceh Utara	The Mean of physical function: 55.67% The Mean Of emotional function: 69.51% The Mean Of social function: 79.02% The Mean of school function: 36.96% Average - average quality of life: 60.48%	The highest mean is a social function, and the lowest mean is a school function