UALITY OF LIFE OF CHILDREN WITH THALASEMIA IN INDONESIA

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QUALITY OF LIFE OF CHILDREN WITH THALASEMIA IN INDONESIA

Literatur Review

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***Abstract*—** Thalassemia is an inherited disorder of hemoglobin (Hb) synthesis, particularly globin chains. 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. Data regarding the condition of children with thalassemia major in Indonesia has not been widely published, but various studies clearly show the high incidence of depression, anxiety and other psychosocial disorders which greatly affect the quality of life of children with thalassemia. Blood transfusions and lifelong use of drugs often lead to feelings of boredom, boredom of treatment, not to mention physical changes, feeling different from relatives or friends will cause feelings of inferiority. They often drop out of school and do not find work, which causes very severe psychosocial effects. Child health servi need to make efforts to optimize children's functional abilities and quality of life so that dren with thalassemia can grow into productive adults. *Pediatric Quality of Life Inventory TM* (sQLTM) can be used to assess a child's quality of life over time. This paper aims to provide an overview of the quality of life of children with thalassemia in Indonesia, so that it can be used s a reference in providing appropriate management regarding the quality of life of children with thalassemia. This literature search uses google scholar, PubMed and ProQuest for complete publications in 2015-2020 with the keywords "quality of life", "children" and halassemia". The inclusion criteria set were the measurement of quality of life using the instrument *Pediatric Quality of Life Inventory* TM (PedsQLTM) in children aged 2-18 years suffering from thalassemia in various regions in Indonesia. ere were 23 research articles related to this search. The average quality of life of children with thalassemia in Indonesia ranges from 50% - 67.2%. Of the four domains essed, school function had the lowest average, followed by emotional function, physical function and social function

Keywords*—*Quality of Life*, *Children*, *Thalassemia*, *Indonesia

INTRODUCTION (*HEADING 1*)

Thalassemia is an inherited disorder of hemoglobin (Hb) synthesis, particularly globin chains. This genetic disease has the most types and frequency in the world. The clinical manifestations vary from asymptomatic 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 a accompanied by severe hemoglobin abnormalities, and 50,000 to 100,000 children die from thalassemia β; 8 of this amount comes from developing countries (1–3).Indonesia is o of the countries in the world thalassemia belt, namely a country with a high frequency of thalassemia genes (number of carriers). This is evident from epidemiological studies in Indonesia which found that the frequency of the beta thalassemia gene ranges from 3-10%. Data obtained from all teaching hospitals in fact 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 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 (2,3).

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. The need for 1 thalassemia major child with a body weight of 20 kg for blood transfusion and adequate iron chelation will cost

around Rp. 300 million per year. This amount does not include the costs of laboratory examinations and monitoring, as well as management of complications that arise (2).

Data regarding the condition of children with thalassemia major in Indonesia has not been widely published, the high incidence of depression, anxiety and other psychosocial disorders which greatly affect the quality of life of children with thalassemia. In Indonesia, a 2009 study on adolescents aged 13-18 years with thalassemia major found that 50.5% of adolescents have a poor quality of life (2,5).

Blood transfusions as a result of the incurable medical condition thalassemia, repeated transfusions. which is tiring and traumatic, complications of disease, and limitations to daily activities at school, play area, or work place and lifelong use of drugs often lead to feelings of boredom, boredom of treatment, not to mention physical changes, feeling different from relatives or friends will cause feelings of inferiority. They often drop out of school and do not find work, which causes very severe psychosocial effects.

Child health services need to make efforts 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.

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 uses google scholar for complete publications in 2015-2020 with the keywords "quality of life", "children" and "thalassemia". The inclusion criteria set were the measurement of quality of life using theinstrument *Pediatric Quality of Life Inventory* TM (PedsQLTM) in children aged 2-18 years suffering from thalassemia in various regions in Indonesia. There were 23 research articles related to this search.

Pediatric Quality of Life Inventory is an instrument that can be used to assess children's quality of life which consists of 23 questions to measure 4 dimensional scales, namely physical function consisting of 8 questions, emotional function 5 questions, emotional function 5 questions and school function 5 questions. The higher the value obtained is considered to have a better quality of life.

RESULT

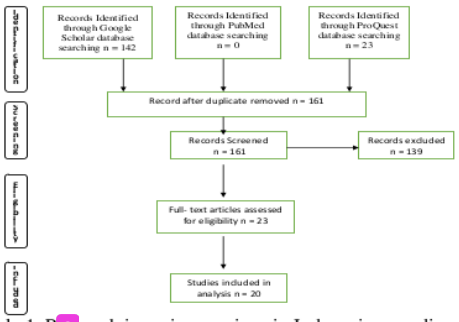

Table 1. Research in various regions in Indonesia regarding the quality of life of children with thalassemia

No	Title (years)	Method	Results	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 of quality of life: 50.9%	The highest mean is social function and the lowest mean is 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 social function and the lowest mean is school function
3	The effect of the thalassemia education package on the quality of life of thalassemia children	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	The highest mean is social function and the lowest mean is school function

	(2014). Hastuti.		function: 79.29% The mean of school function: 53.9% The mean of quality of life: 65.36%	
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 of quality of life: 50.9%	The highest mean is social function and the lowest mean is 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 of quality of life: 62.75%	The highest mean is social function and the lowest mean is 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	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 -	The highest mean is social function and the lowest mean is school function
			(2018) Nikmah and Mauliza.	average quality of life: 60.48%

DISCUSSION

Physical functions that are included in the domain of quality of life assessment illustrate how the child's ability to carry out their daily activities independently.(6) 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 the child's ability to get along with friends at school.(7,8) Assessment of school functions describes a child's ability to do the tasks given at school (9,10).

The results of the 6 studies above can be concluded that the social function of children has the highest average of the other three functions and school function has the lowest average of the other three functions.

School function is the lowest domain in assessing quality of life. This is due to obstruction of school activities and decreased academic achievement scores due to the reasons for the treatment and treatment of thalassemia (9,11–13). This education problem needs to pay attention to 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, the child's susceptibility to disease, and the school is expected to support the treatment of these patients.(13)

Emotional function is the second function after school function. Disturbance of the emotional domain is probably caused by the physical condition of the thalassemia sufferer which causes its own burden (stress condition). This can affect immunological psychoneuro. So if the physical condition is considered as stress, there will be a substance that resembles *beta carboline*, which is a GABA antagonist which is thought to cause a decrease in the number (down regulate) of GABA receptors causing reduced resistance to anxiety and easingreactions *stress* (13). Emotional dysfunction is influenced by various things, namely feeling depressed during diagnosis, therapy that must be undertaken every month regularly, and having to skip school because of having to undergo therapy (9). 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 (14,15). Psychosocial support from families is expected to reduce emotional problems in patients with beta thalassemia major, further explained that psychosocial

support reduces emotional distress, increases the effectiveness of iron chelation and strengthens coping strategies to be better in everyday life (16–18).

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 deposition of iron in the organs.(14,19) Physical changes that 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, refusal to socialize, and going to school (9).

Social function has the highest average score between 60 - 80% among physical, emotional and school functions. This illustrates that the social function of children suffering from thalassemia is still quite good compared to the other three functions. Children can still socialize and adapt to peers or friends at school with all their limitations, the physical changes that occur as a result of treatment and in the limitations of daily activities. Limited physical activity makes the subject unable to do things that can be done by healthy peers (9)

CONCLUSION

The Mean of quality of life of children with thalassemia in Indonesia ranges from 50% - 67.2%. Of the four domains assessed, school function had the lowest mean, followed by emotional function, physical function and social function. Health services that can improve the quality of life of thalassemia patients can 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 is not so much different from the quality of life for normal children.

ACKNOWLEDGMENT *(Heading 5)*

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