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by Yuliani Ayu

Submission date: 08-Apr-2022 08:57PM (UTC+0700)
Submission ID: 1805286123
File name: manuscrift_Ayu_Yuliani-Aris-B-Ing.pdf (164.15K)
Word count: 3018
Character count: 16055

RELATIONSHIP BETWEEN BLOOD TRANSFUSION AND SLEEP DISORDERS IN CHILDREN WITH *THALASSEMIA* IN RSUD '45 KUNINGAN

Yuliani Ayu [1], Aris Diana[2]

1. Lecturer at Tasikmalaya Nursing Department of Tasikmalaya Health Polytechnic
2. A college student at Tasikmalaya Nursing Department of Tasikmalaya Health Polytechnic

Abstract

Thalassemia is a blood disorder disease caused by genetic factors and causes the protein in red blood cells, called hemoglobin, to not function normally, with this condition the child is most likely to have problems, including sleep disorders. Sleep disorders are a collection of conditions characterized by disturbances in the amount, quality, or timing of sleep in an individual. One of the methods used to screen for sleep disorders is using the SDSC (*Sleep Disturbances Scale for Children*). The purpose of this study was to determine the relationship between blood transfusion and sleep disorders in children with *thalassemia*. This study uses *a quasi-experimental* design with a pre-post-test design in one group *(One group pre-post-test design)*. The research sample was 40 respondents using the *total sampling method,* univariat analysis in the form of frequency distribution, and bivariate analysis in the form of *paired sample t-test*. The results showed that there was a significant relationship between the pre-blood transfusion sleep disturbance scale (20%) and the post-blood transfusion sleep disturbance scale (0%) with a p-value of 0.000 and at-count of 4.687. Researchers suggest that parents of respondents understand the importance of blood transfusion for children with *thalassemia*.

Keywords: *thalassemia*, blood transfusion, sleep disorders

Preliminary

Thalassemia is a group of inherited hematologic disorders due to defects in the synthesis of one or more globin chains. Alpha *thalassemia* is caused by a lack or absence of alpha-globin chain synthesis and beta-*thalassemia* is caused by a lack or absence of beta-globin chain synthesis. The imbalance of the globin chains causes hemolysis [1] . Carriers of both alpha and beta-*thalassemia are asymptomatic and do not require therapy.* Patients with beta-*thalassemia* major are at risk of dying from cardiac complications due to iron overload[1,] usually, patients with *thalassemia* require

supportive therapy to maintain their condition. Supportive therapy is carried out to maintain adequate Hb levels to prevent bone marrow expansion and bone deformities and to provide sufficient erythrocytes to support normal growth and activity. One of the supportive therapies for *thalassemia patients* is blood transfusion, which is the basis of the medical management of *thalassemia*[2,3].

Thalassemia cases in Indonesia, thalassemia is the most common genetic disorder. At the *thalassemia center,* Department of Pediatrics (IKA) Faculty of Medicine, University of Indonesia (FKUI) Cipto Mangunkusumo Hospital (RSCM) until the end of 2008 there were 1,455 patients registered consisting of 50% -B *thalassemia*, 48.2% -B/Hb *thalassemia* -E and 1.8% of patients with *thalassemia* -A. It is estimated that every year in Indonesia 2,500 children are born with *thalassemia* (Aji et al, 2009).

This blood disorder causes red blood cells (hemoglobin) to quickly disintegrate so that the lifespan of blood cells becomes shorter and the body lacks blood. For example, if the red blood cells in healthy people can last up to 120 days, in people with thalassemia the red blood cells only last 20-30 days. This disease appears with symptoms including anemia, pallor, difficulty sleeping, weakness, and no appetite. Other complications that can occur in children with thalassemia are swelling of the liver and thinning of the bone marrow [4]. Swelling of the liver and due to severe and prolonged anemia, heart failure is common. Repeated blood transfusions in the hemolysis process (rupture of the hemolysis membrane) cause iron levels in the blood to be very high, so that it is stored in various body tissues such as the liver, spleen, skin, heart, etc. This can result in impaired function of the device (homochromatic)5.

Based on the World Health Organization (WHO) 1993, 4.5% of the total world population is a disease carrier (heterozygous form), which is no less than 250 million people in the world. Of this number, 80-90 million are carriers of thalassemia traits and the rest are carriers of thalassemia traits, other types of carriers of variant hemoglobin traits such as HbE, HbS, HBO, and others6. Now carriers of thalassemia traits reach 7% of the total world population, about 300,000-500,000 babies are born with this disorder. Research by Tarasiuk et al, (2003) stated that children and adolescents with thalassemia have sleep function disorders caused by *Periodic Limb Movements* [4]. In addition, the results of research by Roohangiz et al, (2010) in patients with thalassemia

major found the presence of Restless Legs Syndrome. The incidence of Restless Legs Syndrome is not related to ferritin in levels and iron levels in the body of children with [7] thalassemia

Research Methodology

This study used *a quasi-experimental* design with a pre-posttest design in one group *(One group pre-post-test design).* to know the relationship between blood transfusions and sleep disorders in children with *thalassemia* in the pediatric care room at 45 Kuningan Hospital. The research sample is the object under study and is considered to represent the entire population (Notoatmodjo, 2010). The sample size in this study amounted to 40 children with the criteria of all children with *thalassemia* being treated at RSUD 45 Kuningan and parents allowing their children to be respondents. The instrument used was the SDSC (*Sleep Disturbances Scale Children*) questionnaire consisting of 26 questions about the quality of children's sleep and the format for assessing pulse, temperature, respiratory rate before and after blood transfusion, and blood products used during the study. Previously, the researchers conducted an *Interrater Observer Reliability* test so that the observer's and observer's assessments were the same. In the results of the *Interrater Observer Reliability test* between the researcher and the research assistant, it was found that the Kappa value was 1.0 (Kappa value > 0.8), it was stated that the researcher and research assistant had the same Interrater test value so that the ability between the researcher and research assistant was the same.

Results

The results of this study used univariat analysis that described the characteristics of the respondents including the age of the child and the sex of the child. Bivariate analysis was used to determine the relationship between blood transfusion and sleep disorders in children with *thalassemia* with the following results:

Table 1. Distribution of respondent characteristics based on respondent's age

Characteristics of Respondents	f	(%)
Respondent Age		
Under 3 years (1-3 years)	10	25%
Preschool (4-5 years)	5	12.5%
Children (6-12 years)	22	55%
Teenagers (13-18 years)	3	7.5%
Total	40	100%

Based on table 1, it is found that the distribution of respondents' characteristics by age is mostly at the age of children (6-12 years) which is 55%.

Table 2. Distribution of respondent characteristics by gender

Characteristics of Respondents	f	(%)
Respondent's Gender		
Man	24	60%
Woman	16	40%
Total	40	100%

Based on table 2, it is found that the distribution of respondents' characteristics by gender is mostly male, namely 60%, and female is 40%.

Table 3. Distribution of pulse measurement results between Pre and Post Blood Transfusion

Pulse Measurement Results	Pre Blood Transfusion		Post Blood Transfusion	
	f	(%)	f	(%)
Tachycardia	1	2.5%	2	5%
Normal	37	92.5%	38	95%
Bradycardia	2	5%	0	0%
Total	40	100%	40	100%

Based on table 3 the distribution of pulse measurement results between pre and post-blood transfusions, it was found that before blood transfusion those who had 2.5%

tachycardia were normal. 92.5%, 5% bradycardia. As for the time after the blood transfusion, he experienced 5% tachycardia, 95% normal, and 0% bradycardia.

Table 4. Distribution of temperature measurement results between Pre and Post Blood Transfusion

Temperature Measurement Results	Pre Blood Transfusion		Post Blood Transfusion	
	f	(%)	f	(%)
Hypothermia	28	70%	19	47.5%
Normal	12	30%	19	47.5%
Hyperthermia	0	0%	2	5%
Total	40	100%	40	100%

Based on table 4 the distribution of temperature measurement results between Pre and Post Blood Transfusion, it was found that before blood transfusion, those who had 70% hypothermia were normal. 30%, hyperthermia 0%. Meanwhile, after blood transfusion, 47.5% hypothermia, 47.5% normal, and 5% hyperthermia were performed.

Table 5. Distribution of respiratory measurement results between Pre and Post Blood Transfusion

Breathing Results	Pre Blood Transfusion		Post Blood Transfusion	
	f	(%)	f	(%)
Tachypnea	4	10%	4	10%
Normal	34	85%	36	90%
Bradypnea	2	5%	0	0%
Total	40	100%	40	100%

Based on table 5 the distribution of pulse measurement results between Pre and Post Blood Transfusion, it was found that before blood transfusion those who had 10% tachypnea were normal. 85%, bradypnea 5%. As for the time after the blood transfusion, he had 10% tachypnea, 90% normal, and 0% bradypnea.

Table 6. Distribution of Use of Blood Products for Blood Transfusion

Blood Products	Pre Blood Transfusion	
	f	(%)
Whole Blood (WB)	0	0%
Packed Red Cell (PRC)	40	100%
Total	40	100%

Based on table 6 the use of blood products used for blood transfusions, all respondents use *Packed Red Cell* (PRC) as many as 40 people (100%) or it can be said that all respondents use PRC

Table 7. Distribution of sleep disturbances between Pre and Post Blood Transfusion

Sleep Disorders Scale	Pre Blood Transfusion		Post Blood Transfusion	
	f	(%)	f	(%)
No sleep disturbance	15	37.5%	25	62.5%
Borderline	17	42.5%	15	37.5%
Sleep disturbance	8	20%	0	0%
Total	40	100%	40	100%

Based on table 7 the distribution of sleep disturbances between pre and post-blood transfusions, it was found that before blood transfusions, 37.5% had no sleep disturbances, *42.5* % borderline, and 20% had sleep disturbances. Meanwhile, after blood transfusion, 62.5% had no sleep disturbances, *37.5* % borderline, and 0% sleep disturbances.

The results of a bivariate analysis to determine the relationship between blood transfusions with sleep disorders in children with *thalassemia*. Testing this hypothesis is done by looking at the difference in the scale of sleep disturbances before and after blood transfusion. The difference in the average difference can be seen from the test as follows:

Table 8. T-Test Results of Pre and Post Blood Transfusion Sleep Disorders

Sleep Disorders Scale Comparison	t value	df	*p*-Value
Pre Blood Transfusion-Post Blood Transfusion	4,867	39	0.000

Statistical testing resulted in an at-value of 4.867 with a *p*-value (significance) of 0.000. The value of the t-table for testing with a value of = 0.05 and degrees of freedom Df = 39 is 1.684, so it can be seen that t-count > t-table (4.867 > 1.684) or *p*-value < (0.000 < 0.05), then the hypothesis of a relationship between blood transfusion and sleep disorders in children with *thalassemia* in RSUD 45 Kuningan is acceptable so that it can be seen that there is a significant relationship between the Pre-Sleep Disorder Scale Blood transfusion with sleep disturbance scale Post blood transfusion and it can be concluded that the sleep of children with *thalassemia* is better when after blood transfusion.

discussion

The results of the research on the relationship between blood transfusions and sleep disorders in children with *thalassemia patients* at the 45 Kuningan Hospital found that most children with *thalassemia* were aged children (6-12 years) as many as 22 children or with a percentage of 55%. Clinical symptoms of *thalassemia* have been seen at the age of 2 years, but patients with *Thalassemia* only started treatment at the age of 4-6 years because it was getting paler, resulting in the sufferer requiring periodic transfusions. [8] The results of the research conducted by the researcher are supported by his research by Lazuana T (2014) which states that the proportion of *thalassemia sufferers* who are treated at H. Adam Malik Hospital Medan Untuk Medan To perform transfusion is the most in the age group 15 years as many as 95 people (84%).

The results of the study based on gender found that the highest percentage occurred in men as many as 24 people (60%) compared to 16 women (40%). T *thalassemia* is a

genetic disease caused by a single autosomal recessive allele factor, not a genetic disease caused by an allele factor linked to sex/sex chromosomes, which states that based on gender, 51.6% of *thalassemia sufferers* are male and 48, 4% are female. The number of male patients is more than that of female patients [5].

The results obtained from measurements of pulse, temperature, and respiration showed that most of the respondents experienced slight changes in pulse, temperature, and respiration after blood transfusion but were still within normal limits. This is probably the result of blood transfusions that have been done to maintain hemoglobin levels. Hemoglobin carries 97% of the oxygen that has diffused to the tissues, therefore the oxygen needs in the body can be met so that there is an increase in metabolism in the body which increases pulse, temperature, and respiration to meet the body's needs.

The results of the observation of the use of blood products used for blood transfusions, all respondents used PRC (*Packed Red Cell*). The use of PRC is caused by *thalassemia patients* experiencing abnormalities or deficiency of the hemoglobin chain. This abnormality causes damage to red blood cells in blood vessels so that the lifespan of erythrocytes is short (less than 120 days. Kebutuhan arah pada satu pasien dengan pasien yang lain berbeda tergantung pada hemoglobin pra transfusi yang diperiksa saat akan menjalani transfusi darah. Transfusi darah diberikan pada pasien dengan kadar Hb ≤ 10 g/dL) . Giving blood in the form of PRC 3 ml/kg BW for every increase in Hb 1 g/dL (Permono B, 2006). Children with *thalassemia* do blood transfusions to maintain hemoglobin levels so that anemia does not occur, one PRC bag (150-300 ml) consists of 100-200 ml erythrocytes. This blood product is used in conditions that require the addition of red blood cells only [10].

The results of the research that have been conducted showed that children with *thalassemia before Borderline* blood transfusion were 17 people (42.5%), sleep disturbances were 8 (20%), whereas after *Borderline blood transfusion* 15 people (37.5%), had sleep disturbances. sleep 0 people (0%). It can be seen that there are children with *thalassemia* who experience sleep disturbances before performing blood transfusions. The results of the research conducted by Iqbal (2014) regarding the description of sleep disorders in children with *thalassemia* in the Central Installation of *Thalassemia* and *Hemophilia* RSUD dr. Zainoel Abidin Banda Aceh states that children

with *thalassemia* who have sleep disorders are 9.7%, *borderline* 35.5%, and not sleeping disorders 54.8%.

Children with *thalassemia* before doing blood transfusion look tired and tired. One of the causes of sleep disturbances is fatigue, fatigue also affects a person's sleep patterns. The more tired a person is the shorter the first REM (paradoxical) sleep period. The condition of *thalassemic* children who are tired due to a lack of hemoglobin in the blood may be the cause of sleep disturbances that occur before blood transfusions [12].

The condition of a child with *thalassemia* who has undergone a blood transfusion using PRC is possible that the hemoglobin level in the blood can be fulfilled so that oxygen can be distributed to all parts of the body marked by pulse, temperature, breathing within normal limits, this situation supports the child to sleep better.

The results of the study that analyzed the bivariate variable between blood transfusion and sleep disorders in children with *thalassemia* found that there was a relationship between blood transfusions and sleep disorders in *thalassemic children* with a t-value of 4.867 with a *p*-value (significance) of 0.000, hence the hypothesis of a blood transfusion relationship. with sleep disorders in children with *thalassemia* at the 45 Kuningan Hospital is acceptable. The existence of a relationship between blood transfusion and sleep disorders in *thalassemic children* may be caused by the use of PRC blood products at the time of blood transfusion, slight changes in pulse, temperature, breathing within normal limits after blood transfusion so that children with *thalassemia* can not experience sleep disorders after performing a blood transfusion.

Conclusion

Thalassemia is a group of inherited hematological disorders due to defects in the synthesis of one or more globin chains, usually, *thalassemia sufferers* require supportive therapy to maintain their condition. One of the supportive therapies for *thalassemia patients* is blood transfusion, which is the basis of the medical management of *thalassemia*. Blood transfusion is the process of transferring blood or blood-based products from one person to the circulatory system of another. The results of the study showed that children with *thalassemia* who did blood transfusions at RSUD 45 Kuningan were mostly aged 6-12 years, most were male, there was a slight change

in the measurement of pulse, temperature, respiration between before and after blood transfusion. Children who have not had blood transfusions have sleep disturbances so while children who have undergone blood transfusions sleep better, there is a relationship between blood transfusions and sleep disorders in children with *thalassemia*.

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