



## Family Empowerment Program to Improve Quality of Life in Thalassemia Patient: A Case Study

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### ABSTRACT

**Introduction:** Thalassemia can cause major health problems in children and adversely affect their quality of life, which includes reduced physical, psychological, social, and educational functioning. A family empowerment program is a process or effort to increase the knowledge and willingness of families to maintain and improve the Quality Of Life of children with thalassemia

**Objective:** The purpose of this case study is to carry out nursing care for children with thalassemia and quality of life problems with the Family Empowerment Program (FEP) approach.

**Method:** The method used is descriptive-qualitative with a case study approach. As well as pre-test intervention and post-test intervention. Nursing care was carried out with the author for three days and the family intervened using the Family Empowerment Program (FEP) approach for three weeks. The subject of this case study is a thalassemia child with poor quality of life. Data collection techniques include interviews, observation, physical examination, and documentation.

**Results:** The results of the case study at the assessment stage found that An.S. had problems in the dimensions of social function and school function due to complex or long-term treatment or treatment programs and unsatisfactory interpersonal interactions. Intervention and implementation used in An.S. are using family empowerment with the Family Empowerment Program (FEP) approach.

**Conclusion:** The conclusion of nursing care in children with thalassemia was that during the intervention of three meetings, there were few changes that were positive for the family and the client in handling social function and school function. After the intervention of the Family Empowerment Program (FEP) approach for three weeks, the results of filling out the PedsQL questionnaire show that the dimensions of physical function are good (90.6), the dimensions of emotional function are good (95), the dimensions of social function are good (80) and the dimensions of school function are good (85).

**Keywords:** family empowerment program, thalassemia, quality of life

## Introduction

Thalassemia is the world's most prevalent hereditary blood disease or disorder, defined by the lack of synthesis of one of the polypeptide chains that can affect the rate of hemoglobin formation, resulting in clinical signs of severe anemia (Kamil et al., 2020)(Astarani & Siburian, 2016). Thalassemia can be found worldwide, with some tropical countries having the highest prevalence of the thalassemia gene. Approximately 3% of the world's population has the thalassemia gene with the highest incidence of up to 40% of cases in Asia. In the Southeast Asia region, there are 55 million thalassemia carriers (Marnis et al., 2018).

Thalassemia is classified into two types: thalassemia minor and thalassemia major. The difference between the two is that people with mild thalassemia (minor) can live a normal life and do not require special therapy or medication (Wacharasin et al., 2015b). However, people with thalassemia minor can pass on thalassemia disease to their offspring. Whereas someone with thalassemia major requires special care and therapy because thalassemia major is a severe blood disorder (Yanitawati et al., 2017). Patients with thalassemia major are unable to produce enough red blood cells and will almost certainly need blood transfusions throughout their lives.

Thalassemia has exacerbated numerous global health issues, especially in developing countries, prompting the World Health Organization to launch a program to address it (Supartini et al., 2013). According to the World Health Organization (WHO), 7% of the world's population carries the characteristics of thalassemia. Every year, between 300,000 and 500,000 neonates are born with severe hemoglobin disorders, and between 50,000 and 100,000 children die from thalassemia (Kamil et al., 2020).

The highest prevalence of thalassemia is in China, the Middle East, the Mediterranean, the Pacific Islands and in South Asia, and is growing rapidly in many continents, including Europe, the Americas and Australia (Wacharasin et al., 2015a). According to the Thalassemia Foundation of Indonesia, the number of thalassemia cases is increasing. In 2012, there were 4,896 thalassemia cases until June 2021 when there was an increase in thalassemia cases to 10,973 cases and Indonesian epidemiology states that the frequency of the Thalassemia gene ranges from 3-10%. The number of thalassemia cases in West Java is 35% (Listyaningsih et al., 2023).

Thalassemia can cause major health problems in children and adversely affect their Quality Of Life, which includes reduced physical, psychological, social, and educational functioning (Pranajaya & Nurchairina, 2017). A person with thalassemia experiences severe anemia, pain, physical abnormalities (enlarged liver and spleen), thalassemia-specific facial features (Cooley facies), delayed or arrested growth and puberty, short stature and severe complications such as heart problems, liver problems, endocrine problems and infections (Listyaningsih et al., 2023).

Thalassemia is a chronic anemia disease that requires children to receive supportive therapy in the form of blood transfusions and lifelong chelation therapy. This can lead to iron accumulation in the body (hemosiderosis) (Yanitawati et al., 2017). Hemosiderosis is a disorder in which blood ferritin levels exceed 1,000 mcg/l, causing heart, liver, and metabolic hormone problems, as well as other significant consequences that can lead to death. The body cannot produce iron physiologically, so iron chelation therapy is needed to remove accumulated iron in the body, and to avoid long-term complications and abnormalities.

However, the administration of such therapy may cause certain health problems (Borimnejad et al., 2018).

Apart from physical problems, a child with thalassaemia often faces psychosocial problems as well as functioning problems in school. Among the psychological problems faced are self-concept issues and stigma (Borimnejad et al., 2018). Children with thalassaemia will feel stress due to the long medical treatment. Children with thalassaemia have less opportunity to play, attend school, and socialize with their classmates; physical and psychological difficulties may interfere with their Quality Of Life. As a result, it is imperative to focus on the areas of Quality Of Life of thalassaemia children (Astarani & Siburian, 2016).

The role of parents is needed in supporting and paying attention to children's health with changes in Quality Of Life in children with thalassaemia. Parents need to provide support and provide effective coping strategies so that children and parents are able to deal with chronic diseases experienced by children so that they can adapt positively and can improve the Quality of Life of children with thalassaemia (Marnis et al., 2018).

Efforts to improve the Quality Of Life of thalassaemia patients require parental support in the form of family empowerment. Family empowerment program is a process or effort to increase the knowledge and willingness of families to maintain and improve the Quality Of Life of children with thalassaemia (Borimnejad et al., 2018). It is intended that families provide support and take steps to maintain and improve their health and quality of life through the family empowerment program (FEP) which is a persuasive effort. Changes or actions in providing support, as well as maintaining and improving health, are formed through a learning process, so that habits are expected to last long and last because they are influenced by awareness. (Yanitawati et al., 2017).

The family's capacity to care for their family members shows a form of family empowerment to provide assistance to patients and can have a good influence on health and Quality Of Life in children with thalassaemia. Research on family empowerment programs was conducted by (LM Widyastuti, 2020) which showed that there was an influence of family empowerment programs on the Quality Of Life of children with thalassaemia.

According to the findings of research conducted by (Borimnejad et al., 2018), Quality Of Life of children who vary results in normal life outcomes in 21 (84%) respondents and Quality Of Life at risk in 4 (16%) respondents. The results of the chi-square test obtained ( $=0.001$ )  $<0.05$ , this shows that there is an influence between parental support and the quality of life of children with thalassaemia in the Pediatric Poly Room Dr. Soeroto Ngawi. This contradicts research conducted by (Listyaningsih et al., 2023) which says that there is no influence before and after FEP on Quality of Life of thalassaemia children at RSUD dr. Soediran Mangun Sumarso.

## **Objective**

The purpose of this case study is to gain real experience and be able to carry out nursing care for children with thalassaemia with Quality Of Life problems with the Family Empowerment Program (FEP) approach.

## **Method**

The design used by the author is to use a case study design with a nursing care approach. This case study uses an analytic descriptive method, which is with the main objective to explore the problem, provide an overview of the case study and analyze more deeply about

nursing care with the Family Empowerment Program (FEP) approach to Quality Of Life in children with thalassemia. Pre test and post test intervention were conducted. Post Test Intervention is carried out after three weeks of application of the Family Empowerment Program (FEP) approach. The case study framework can be seen below:

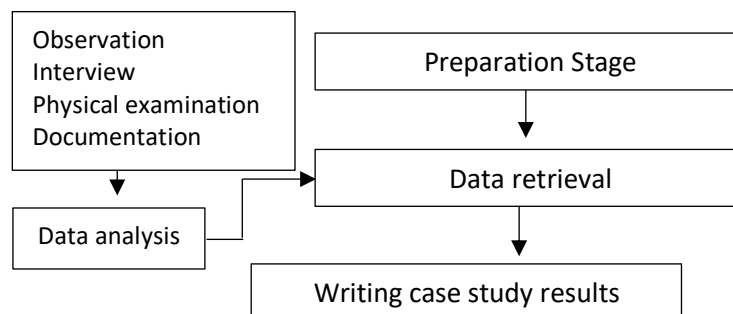


Figure 1. Case Study Framework

The subjects in this case study were clients with thalassemia according to the inclusion and exclusion criteria. Inclusion criteria included clients with thalassemia, receiving treatment in the hospital by undergoing blood transfusions, able to communicate verbally (interacting cooperatively), willing to be a study subject and filling out informed consent. Exclusion criteria in this case study are clients with thalassemia who are treated intensively during the study and who are currently hospitalized with diseases other than thalassemia. Data collection includes interviews, observation, physical examination and documentation while still using ethics in research (informed consent, anonymity, confidentiality, beneficence, voluntary, autonomy, justice).

The tools used in this case study are a set of physical examination tools, all tools used in nursing care actions, and questionnaires. The questionnaire used is the Pediatric of Life Inventory (PedsQL) instrument which consists of four dimensions, namely the dimensions of physical, emotional, social and school function dimensions. In giving values if you never experience a value of 0, almost never experience a value of 1, sometimes a value of 2, often a value of 3 and almost always given a value of 4. The value is interpreted as follows: if the value < 77.0 is categorized as poor Quality Of Life and if the value > 77.0 is categorized as good Quality Of Life.

## Result

### Assessment

The client named An.S aged 13 years old from Kampung Rawabalah, Pusakajaya Village, Pasirkuda District, Cianjur Regency with thalassemia. The client was diagnosed with Thalassaemia at the age of three months. Prior to diagnosis, the client appeared pale and weak, with no bleeding, body heat, or bruising. He was then transferred to Pagelaran Cianjur Hospital for treatment due to his complaints of shortness of breath and seizures. She was diagnosed with Thalassemia after a hemoglobin test showed that her Hb was 6.9 gr/dl. The client underwent a blood transfusion. After that, the client underwent routine blood transfusions. According to the client's mother's statement, the client had received basic vaccines. Only the measles vaccination was missed due to the illness.

At the time of assessment, it was found that the general condition was good, composmentis consciousness, the client looked pale, complained of slight weakness. Blood pressure: 90/60 mmhg, respiration: 24X/min, pulse: 90x/min and temperature: 36.5°C, the abdomen is slightly bulging, there is blackness around the knees, black spots on both legs and the fingernails look pale. The client also said that the client had difficulty in getting along with friends his age, other children rarely wanted to be friends with him, were often made fun of by his friends and felt different from other friends because he had to go to the hospital to do tranfsusion. Hasil pemeriksaan atau pengisian kuesioner pada klien dengan menggunakan kuesioner PedsQL dan didapatkan hasil sebagai berikut :

Table 1. PedsQL quality of life measurement before FEP intervention

<b>Dimensions</b>	<b>Score</b>	<b>Quality Of Life</b>
Physical function	87.5	Good
Emotional function	90.0	Good
Social function	50.0	Poor
School function	60.0	Poor
Average Quality Of Life	71.875	Poor

Interpretation:

Score < 77.0: poor

Score > 77.0 : good

### Analysis

The results of the assessment obtained nursing problems that arise are poor quality of life in An.S, can be seen in the results of filling out the PedsQL questionnaire which shows that in the dimension of social function with a value of 50 (poor) and in the dimension of school function with a value of 60 (poor). With an average quality of life value in An.S of 71.875 (less than 77.0) with the interpretation that An.S has a poor Quality Of Life.

### Diagnosis

The results of the assessment obtained nursing problems that arise in poor Quality Of Life in An.S, with problems in the dimensions of social function and school function due to complex or long-term care / treatment programs and unsatisfactory interpersonal interactions.

### ***Intervention***

Nursing interventions and activities need to be established to increase, improve, and maintain An.S's effective Quality Of Life in clients with family empowerment using the family empowerment program (FEP) approach.

### ***Implementation***

In this implementation there are procedures carried out to clients for the improvement, improvement, and maintenance of An.S effective Quality Of Life in clients with family empowerment using the family empowerment program (FEP) approach. The implementation of nursing care is carried out for three meetings by the nurse and interventions are carried out by the family for three weeks to see changes and their effect on Quality Of Life.

### ***Evaluation***

During the intervention of three meetings there were few changes that were positive towards the family and client in handling social function and school function in An.S. Likewise, after the intervention the family was involved as a family empowerment with the Family Empowerment Program (FEP) approach for three weeks, with the results of filling out the PedsQL questionnaire which showed that in the dimension of physical function with a value of 90.6 (good), the dimension of emotional function with a value of 95 (good), the dimension of social function with a value of 80 (good) and in the dimension of school function with a value of 85 (good). Supported by a statement after three weeks of implementing the intervention, the client who said that the client had begun to be able to socialize with friends at school, had started to get along with friends his age, had counted quite a lot of friends who wanted to be friends with him, and had been able to arrange a schedule to the hospital with his school.

### ***Discussion***

According to the research findings on An.S, the client's mother denied that she and her husband had thalassemia or carriers. Blood tests conducted on the client's mother and husband showed negative results. From the results of the assessment, there is a gap in the theory which states that "Thalassemia is the most common hereditary blood disease or disorder in the world, which is determined by the lack of synthesis of one of the polypeptide chains that can affect the rate of hemoglobin formation, resulting in clinical signs of severe anemia" (Kamil et al., 2020). This could be because genetic screening was not carried out on the grandparents of An.S's parents (in the generation before the client's father and mother) and was not proven by physical evidence of the results of the examination carried out by An.S's parents, only denial from her mother.

At the time of the other assessment, it was found that the client looked pale, complained that the body was a little weak, the stomach was slightly distended, there was blackness around the knees, black spots on both legs and the fingernails looked pale. This is in accordance with the theory conveyed by Amelia et al., (2018), which states that clients with thalassemia have a paler appearance, mongoloid face shape (cooley facies), icterus, growth disturbance, splenomegaly and hepatomegaly, which causes an enlarged abdomen,

pathological fractures caused by blood tissue hyperplasia, and malocclusion, which is a consequence of iron accumulation in the skin.

From the physical differences as well as by requiring a person with thalassemia to carry out a complex (long-term) care or treatment program with a healthy child of the same age, which can cause the client to have difficulty in getting along, other children rarely want to be friends with him, are often made fun of by his friends and feel different from other friends.

For children with thalassemia, nursing diagnoses are based on data obtained from research, which consists of nursing problems. Nursing diagnosis is based on the formulation of PES (problem, etiology, and symptom), where the problem or problem is taken from the Indonesian Nursing Diagnosis Standards (SDKI). Whereas in the case of An.S the author focuses on the problem of poor Quality of Life which is not in the Indonesian Nursing Diagnosis Standards book (SDKI). The results of filling out the PedsQL questionnaire obtained nursing problems that arise, namely in the dimensions of social function and school function with a value of less than 77.0 which classifies that social function and school function in An.S are poor.

Nursing care plans include: determining goals, and nursing action plans. The role of parents is needed to support and to be able to pay more attention to children's health to changes in Quality Of Life in children with thalassemia. Parents need to provide support and be able to provide good and effective coping strategies so that children and parents are able to accept and deal with chronic diseases experienced by children so that they can easily adapt positively and can improve the Quality Of Life of children with thalassemia (Marnis et al., 2018).

Efforts to improve the Quality Of Life of thalassemia patients require parental support in the form of family empowerment. Family empowerment program is a process or effort to increase the knowledge and willingness of families to maintain and improve the Quality Of Life of children with thalassemia (Borimnejad et al., 2018). It is intended that families provide support and take steps to maintain and improve their health and quality of life through the family empowerment program (FEP) which is a persuasive effort. Changes or actions in providing support, as well as maintaining and improving health, are formed through a learning process, so that habits are expected to last long and last because they are influenced by awareness (Yanitawati et al., 2017).

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According to the expected outcome criteria, nursing implementation is a series of activities carried out by nurses to help clients experiencing health problems with good health status. The quality of nursing care provided to clients includes support, treatment, improvement of conditions, family education, or prevention of future health problems. The client's needs, other factors that influence nursing needs, implementation methods, and communication activities should be the focus of the implementation process (Pranajaya & Nurchairina, 2017).

In the implementation carried out, the author always refers to all plans that have previously been compiled. In this case the author has identified the expectations of patients and families in achieving life, realizing that the conditions experienced have important value, actively involving patients in care, providing opportunities for patients and families to be

involved with group support with modified Family Empowerment Program (FEP) interventions, recommending expressing feelings about conditions realistically, training to set goals in accordance with expectations and training how to remember achievements, good experiences.

The final stage of the nursing process, often known as nursing evaluation, can determine whether the objectives of nursing actions are achieved or whether additional steps are needed. This stage evaluates the success of the plan and implementation of nursing actions to meet the client's needs (Amelia et al., 2018).

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There are two types of evaluation, namely summative and formative evaluation using several methods (Amelia et al., 2018). Evaluation can be divided into 2 types, namely ongoing evaluation (summative) and final evaluation (formative). Summative evaluation is an evaluation that is done in the form of filling out a progress note format oriented to the problems experienced by the family. the format used is the SOAPIER format. Likewise, what is done by the author in making nursing evaluations is by using the SOAPIER format. While formative evaluation is an evaluation that is done by comparing the goals to be achieved. If there is a gap between the two, perhaps all stages in the nursing process need to be reviewed, in order to obtain data, problems or plans that need to be modified (Amelia et al., 2018).

## **Conclusion**

The conclusion of nursing care in children with thalassemia during the intervention of three meetings there were few changes that were positive towards the family and the client in handling social function and school function. after the intervention of the Family Empowerment Program (FEP) approach for three weeks, with the results of filling out the PedsQL questionnaire which shows that the dimensions of physical function are good (90.6), the dimensions of emotional function are good (95), the dimensions of social function are good (80) and the dimensions of school function are good (85).

## ***Conflict of interest***

There is no conflict of interest.

## ***Ethical approval***

This research has received ethical approval form the Health Reseach Ethic Commission of the STIKes Muhammadiyah Ciamis number 019/KEPK-STIKESMUCIS/XII/2022.



### **Authors' contribution**

Each author contributed equally in all the parts of the research. All authors have critically reviewed and approved the final draft and are responsible for the content and similarity index of the manuscript.

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