Psychological Problems of Pediatric Patients with Thalassemia

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ABSTRACT

Pediatric patients with thalassemia experience various psychosocial problems that have an impact on the decreased quality of life resulting in depression. The purpose of this study was to look at psychosocial problems and things that happen to children and adolescents with thalassemia related psychosocial experiences. The literature study was made by analyzing scientific articles published from 2015-2020 and in English. Data were obtained from the Scopus, Wiley, Pumbed, and Proquest databases with the keywords psychosocial, pediatric, thalassemia, and quality of life. The results of the literature study found 11 articles that matched the inclusion and exclusion criteria. The course of the disease from thalassemia that requires intense treatment, the patient faces a considerable psychological burden. Social isolation, decreased self-esteem, poor academic performance, contribute to psychological stress. This will have an impact on decreasing the quality of life. The results of the review of the literature review can be used as basic data in the development of counseling and mentoring programs for pediatric and adolescent patients with thalassemia major.

Introduction

The most common form of hemolytic anemia is beta thalassemia, and around 60,000 thalassaemic newborns are born each year around the world.1 Thalassemia is a blood disorder that is inherited. Beta-thalassemia is a genetically determined chronic hematological condition characterized by severe hemolytic anemia due to a disorder of synthesis of hemoglobin chains. resulting in variable phenotypes ranging from severe anemia to clinically asymptomatic individuals. Three main forms have been described: thalassemia major, thalassemia intermedia and thalassemia minor. Individuals with thalassemia major usually present within the first two years of life with severe anemia, requiring regular red blood cell (RBC) transfusions.2 Anemia requires frequent blood transfusions to sustain life, whereas hemosiderosis and other illness consequences require a long and distressing treatment program. Thalassemia is a chronic condition with a variety of clinical and psychological complications. Physical deformities, growth retardation, and delayed puberty are all possible consequences of thalassemia. It has a negative impact on physical appearance, such as bone abnormalities and short height, which contributes to a negative self-image.2 Patients with thalassemia frequently experience severe problems such as heart failure, cardiac arrhythmia, liver disease, endocrine issues, and infections. Children with chronic physical illnesses exemplified by thalassemia are vulnerable to emotional and behavioral problems.2,3

The onset of symptoms, the rigors of therapy, and frequent absences from school place a significant strain on the emotional and interpersonal resources of children and their families. Patients with thalassemia frequently experience severe problems such as heart failure, cardiac arrhythmia, liver disease, endocrine issues, and infections. A child with
thalassemia has emotional and cognitive requirements that are vastly different from those of adolescents seeking independence and identity. According to several sources, up to 80% of children with thalassemia are likely to experience psychological issues such as oppositional defiant disorder, anxiety disorders, and depression. Although children with all forms of blood disorders face similar challenges, those with thalassemia are special in that they must visit the hospital on a regular basis for blood transfusion. According to Clemente et al, different blood illnesses have distinct effects on children, resulting in greater incidence of psychological difficulties in children with thalassemia. A study of adolescent with thalassemia's Quality of Life (QOL) found that those with psychiatric symptoms had a lower QOL. Because of the chronic nature of thalassemia and its intense and demanding treatment, patients and their families face a considerable psychological burden. Social isolation, low self-esteem, poor academic performance, and stigmatization all contribute to psychological distress. Patients with thalassemia major require additional psychological care in order to alleviate mental stress and improve their competitiveness. Additional psychological assistance is required for patients with thalassemia major in order to alleviate emotional stress, develop competence, and improve therapeutic compliance in daily life.

**Methods**

This literature study was carried out by summarizing and analyzing articles related to the study questions and objectives. The search method was conducted using several electronic databases, namely Scopus, Wiley, Pubmed with the keywords psychosocial adolescent, thalassemia, and quality of life. Inclusion criteria in this literature search are (1) study articles that have titles and content that are in accordance with the study objectives; (2) full text; (3) in English; (4) quantitative and qualitative study articles; (5) year of publication 2015-2021. While the exclusion criteria are (1) the article does not have a complete structure; (2) in the form of article review. From this search 11 articles were found that matched the inclusion and exclusion criteria.

**Table 1. Article review table**

<table>
<thead>
<tr>
<th>Author</th>
<th>Title</th>
<th>Design</th>
<th>Sample</th>
<th>Data Collection Method</th>
<th>Result</th>
<th>Conclusion</th>
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<tbody>
<tr>
<td>Yasmin and Hasnain, 2018</td>
<td>Quality of Life of Pakistani Children with β-Thalassemia Major</td>
<td>Observational analytic study</td>
<td>200 children, aged 5-25 years old</td>
<td>Using the TranQOL questionnaire to assess physical health, health emotional, function family support, school function and career</td>
<td>The mean value of TranQOL 48.33 ± 5.6, family function 53.86 ± 13.6, function school and career 39.70 ± 18.4. education, age, pain, level ferritin was significantly associated with the value of TranQOL</td>
<td>Family support and strategies for reduce sadness and frustration such as counseling facilities are very necessary to improve the quality of life of children with thalassemia.</td>
</tr>
<tr>
<td>Shafie, Chabra et al, 2020</td>
<td>Health-related quality of life among children with transfusion-dependent Thalassemia (TDT): A cross-sectional study</td>
<td>Cross sectional</td>
<td>368 Malaysian children with TDT</td>
<td>HRQOL survey of Malaysian children with TDT was conducted using the PedsQL™ 4.0 Generic Core Scales</td>
<td>The mean (SD) Psychosocial Health Summary Score (PCHS) was 79.39 (14.81). Predictors for a lower PCHS was the use of dual chelating agents(R^2 = 0.041), F (1,</td>
<td>The HRQOL of TDT children in Malaysia has improved over the last decade owing to the better access in treatment. However, further effort is needed to improve the</td>
</tr>
</tbody>
</table>
Malaysia

Malaysia

36(2) = 15.60, 
p = < 0.001). The school functioning score had the lowest mean (SD) score of 69.52(20.92) in the psychosocial dimension.

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| Boonchoodang N, Louthrenoo, et al, 2015 | Health-Related Quality of Life in Adolescents with Thalassemia | Cross sectional | 64 adolescent patient with thalassemia aged 13-18 years old and also the parent of these 64 patients | The Pediatric Quality of Life Inventory 4.0 Scales (PedsQL 4.0) self-report form was administered to the adolescents in both groups. Parents were also asked to complete the PedsQL 4.0, parent proxy-report form. The self-reported total, psychosocial, and school functioning scores of the thalassemia patients were significantly lower than those of the healthy controls (p = 0.03, 0.04, and <0.001, respectively). The parent-reported psychosocial and school functioning scores of the thalassemia group were also significantly lower than those of the controls (p = 0.03 and 0.003, respectively). Among adolescents with thalassemia, the serum ferritin level and comorbidity were the only variables associated with quality of life scores. | thalassemia negatively affected quality of life. For a better quality of life, thalassemia patients should be monitored for serum ferritin levels and treated for comorbidity as part of their comprehensive health care. |
| Hakeem GLA, Mousa et al, 2018 | Health-related quality of life in pediatric and adolescent patients with transfusion-dependent ß-thalassemia in upper Egypt (single center study) | Case control | 64 patients recruiting pediatric hematology outpatient clinic from July 2014 to February 2017, 8-18 years old | PedsQL™ 4.0 Generic Core Scale (Arabic version) Mean physical, emotional, social, school performance, psychological and total scores (-36.9 ± 20.9, 49.4 ± 17, 47.2 ± 21.3, 38.5 ± 15.5, 45.3 ± 13.8, 47.9 ± 38.8 respectively) were significantly decreased compared with Scheduled programs giving psychosocial help and a network connecting between the patients, school officials, thalassemia caregivers and the physician is required especially in developing countries where the health services are not |
control (p = 0.001 for all). Older age of starting transfusion was statistically significant protecting factor from poor physical QOL in thalassemia patients (OR = 0.96, p = 0.03).

### Mikael NA, Al-Allawi, 2018

**Factors affecting quality of life in children and adolescents with thalassemia in Iraqi Kurdistan.**

**Case control**

100 thalassemic patients and 100 healthy subjects between the ages of 6-18 years were enrolled.

(PedsQL) 4.0 was administered by both child and parent reports.

The mean HRQoL score of thalassemic patients was significantly lower than that of healthy subjects, with lowest scores in physical functioning. Furthermore, the mean HRQoL of TM was significantly lower than that of TI subgroup. Significantly lower mean HRQoL scores were seen in those taking ≥6 transfusions/year, with hepatitis C infection, with illiterate parents, and those on oral iron chelation. Pearson correlation revealed that HRQoL was negatively associated with age, frequency of transfusions, and serum ferritin, but positively correlated with age at starting transfusion and age at diagnosis. Only age and serum ferritin remained significant by multivariate analysis.

**Integrated with social organizations.**

Special school services for thalassemia patients are required to deal with the repeated absence and anemia induced low mental performance of thalassemia children.

This study shows that among Iraqi Kurds with thalassemia, the disease has a significant negative impact on quality of life, with age and serum ferritin being identified as independent predictors. Psychosocial, educational, and patient-centered management programs may be needed to improve HRQoL in this disease.
<table>
<thead>
<tr>
<th>Study</th>
<th>Design</th>
<th>Group 1</th>
<th>Group 2</th>
<th>Analysis</th>
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<tbody>
<tr>
<td>Alhakeim, Najm, 2020</td>
<td>Case control</td>
<td>children (6-12 years)</td>
<td>The Children’s Depression Inventory (CDI), iron status (serum iron, ferritin, transferrin, TS%), and serum levels of CCL11, IL-1β, IL-10, and TNF-α were measured in TDT with (n = 54) and without (n = 57) a major depression-like episode (MDLE) and in healthy children (n = 55).</td>
<td>MDLE due to TDT is associated with a greater number of blood transfusions and increased iron overload and IL-1β levels. Partial least squares path analysis shows that 68.8% of the variance in the CDI score is explained by the number of blood transfusions, iron overload, and increased levels of IL-1β and TNF-α. The latter two cytokines partly mediate the effects of iron overload on the CDI score, while the effects of blood transfusions on the CDI score are partly mediated by iron overload and the path from iron overload to immune activation. Iron overload is also associated with increased IL-10 and lower CCL11 levels, but these alterations are not significantly associated with depression. Blood transfusions may be causally related to MDLE in TDT children and their effects are in part mediated by increased iron overload and the consequent immune-inflammatory response. The results suggest that effects of iron overload and its consequences including inflammation and oxidative stress toxicity may cause MDLE. Current treatment modalities with folic acid and vitamin C are insufficient to attenuate iron overload and immune-inflammatory responses and to prevent MDLE in children with TDT.</td>
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<tr>
<td>Toret, et al, 2018</td>
<td>Cross sectional</td>
<td>57 β-thalassemia major (β-TM) patients and the control group</td>
<td>The short form-36 (SF-36) questionnaire and Beck depression inventory (BDI) were used</td>
<td>The mean SF-36 scores of the patient and the control groups were 59.2 ± 12.4 and 75.7 ± 11.8, and the mean BDI scores of the patients and controls were 13.5 ± 6.4 and 6.1 ± 3.7, respectively. There was a statistically significant difference in QoL between the groups. The β-TM patients have a comparatively worse QoL score than the normal population. Improving QoL should be the target of clinicians who are monitoring adolescent or young adult β-TM patients.</td>
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<tr>
<td>Author(s)</td>
<td>Title</td>
<td>Study Design</td>
<td>Sample Size</td>
<td>Measured Outcomes</td>
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<td>-----------------------------------------------</td>
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<tr>
<td>Abdul, Ahmed, Hassan, 2016</td>
<td>Health-related Quality of Life in Children and Adolescents With β- Thalassemia Major on Different Iron Chelators in Basra, Iraq.</td>
<td>Case control</td>
<td>209 children and adolescents aged 2 to 17 years old</td>
<td>HRQoL was assessed using the Pediatric Quality of Life (PedsQL) Generic Core Scale questionnaire, version 4.0, for children 2 to 12 years old and the Short Form-36 health survey questionnaire, version 2 (SF-36v2), for children and adolescents aged 13 to 17 years old</td>
</tr>
<tr>
<td>Batool et al, 2017</td>
<td>Psychosocial burden among thalassemia major patients: an Explanatory investigation of South Punjab Pakistan</td>
<td>Cross sectional</td>
<td>91 adolescent with thalassemia</td>
<td>structured interview</td>
</tr>
<tr>
<td>Ishfaq et al, 2018</td>
<td>Psychosocial Problems Faced By Thalassemia Major Patients of</td>
<td>Cross sectional</td>
<td>Two hundred thalassemia major patients were</td>
<td>structured interview and analyzed by using SPSS (Statistical Package for the significant association between the education level of thalassemia major patients</td>
</tr>
</tbody>
</table>
Discussion

Based on the search results using these keywords, 11 articles were obtained. The review table can be seen in Table.1. This literature review consists of 11 articles that discuss the psychosocial problems of pediatric patients with thalassemia major, consisting of qualitative and quantitative study articles. There are 2 qualitative study articles and 10 quantitative study articles. Quantitative study articles use questionnaires such as PedsQL, HRQOL, PSCL SDQ, CDI, and TranQOL to assess adolescent quality of life related to adolescent psychosocial thalassemia. The results of the study using the HRQOL questionnaire shows that adolescents with thalassemia have psychosocial disorders that cause a decrease in their quality of life. the HRQoL mean scores across dimensions decrease, except for the emotional dimension, which improved as patients grew older. The scores between the genders were similar across the dimensions. Apart from the emotional dimension, the negative correlation and coefficient between age and the HRQoL scores implies that HRQoL worsens as patients grow older. As patients grow older, the burden of treatment may increase with higher volume of blood required, onset of complications and the need for higher dosages of iron chelating therapy, resulting in lower HRQoL. Understanding how
the disease and treatment affects the HRQoL dimensions is imperative in improving the delivery of care beyond the clinical markers of the disease. It is in line with the results of a study conducted by Behdani et al (2015) using the PSCL questionnaire showed that there was a significant relationship between depression, anxiety, quality of life, and behavioral screening between thalassemia major adolescents and healthy adolescents. Patients with thalassemia have a lower QOL than their peers (P = 0.001), the rate of depression is higher in this group (P = 0.015), Also behavioral problems in these children are more than healthy subjects. In addition, adolescents with thalassemia major have more psychological problems than healthy adolescents, so that the level of depression in adolescents with thalassemia major is also high.

While in a study conducted by Yasmeen and Hasnain (2018) with using the TranQOL questionnaire showed that the TranQOL scores in thalassemia children were 53.86 ± 13.6 for family function, and 39.70 ± 18.4 for school and career functions. In a qualitative study article conducted by Ishfaq, Diah, Ali, Fayyaz, and Batool (2018) with using structured interviews showed that there was a significant relationship between the education level of adolescents with thalassemia major and gender and a significant relationship between adolescent boys and girls with thalassemia major in extracurricular activities at school.

Batool, Ishfaq, and Bajwa (2017) also conducted qualitative study using structured interviews with study results showing that thalassemia has an impact on education, adolescent socialization does not participate in playing outside the home with their peers, adolescents feel dissatisfied with their body image, adolescents feel In contrast to their siblings, adolescents receive the same attention from their parents, adolescents feel a burden to their parents, adolescents feel that thalassemia disease limits their social life, and adolescents feel that there is a high psychosocial burden.

Adolescents with thalassemia major often experience pain and should be given restrictions in terms of activity. This restriction must be done to maintain the condition of adolescents with thalassemia major who are easily tired. This is in line with the results of study conducted by Ishfaq et al (2018) which states that thalassemia major adolescents have difficulty in carrying out extracurricular activities at school. This is because their bodies are weak and tired easily. Therefore they are given restrictions in terms of activities so that their body condition can be stable. The results of study conducted by Batool et al (2017), there are teenagers who say that they don't like to see or watch other people do things they can't do. In addition, thalassemia major adolescents also feel insecure with the difficulty of increasing height, skin that turns black, and changes in facial bones, so this can interfere with body image.

Restrictions on physical activity given to thalassemia major adolescents certainly make adolescents feel sad even frustrated so that this is very disturbing psychologically, and makes thalassemia major adolescents also experience anxiety, decreased self-esteem, and even depression which can have an impact on the quality of life of thalassemia major adolescents.

Physical restrictions also have an impact on the social relationships of thalassemia major adolescents because it makes them isolated from peers and the people around them. Circumstances like this can make teenagers limit their association with their peers. The decline in school performance in adolescents with thalassemia major is due to the fact that adolescents are often absent to get blood transfusions every month at the hospital. This has an impact on low academic achievement, which can make adolescents have great emotional demands.

Psychosocial is part of the quality of life of adolescents with thalassemia major, so that when the psychosocial of adolescents is disturbed, it will affect their quality of life. This is
in line with the results of study conducted by Batool et al (2017) which states that adolescents with thalassemia experience a decreased quality of life with a total HRQOL of 79.50 and lower in female adolescents (75.29). The same thing was also expressed by Yasmeen and Hasnain (2018) that the quality of life of adolescents with thalassemia which includes physical, emotional, social, and school functions is lower than healthy adolescents, especially in the psychosocial domain and school function (p<0.001). This is because starting from the appearance of symptoms of the disease, treatment that lasts a lifetime, and frequent absences from school make teenagers have great emotional demands. The same opinion was also expressed by Karande and Kulkarni (2005) who stated that the decline in school performance in adolescents with thalassemia major was due to the fact that adolescents were often absent from receiving blood transfusions every month at the hospital. Social isolation, decreased self-esteem, low academic achievement, and a bad stigma will lead to the psychological burden of thalassemia adolescents. The need to implement programs that target the patients’ perceived issues relevant to their disease, including psychosocial support to improve the patient self-image and self-esteem, and facilitating normal life style. Moreover, educational programs which take in consideration the education backgrounds of the parents, and centered on handling various complications promptly, safe transfusion practices, and ensuring the regular and correct use of chelators would be instrumental. So that Adolescents with thalassemia major need additional support psychological factors to reduce stress, strengthen competence, and adherence to therapy for daily living.

Conclusion

Psychosocial problems arise in adolescents with thalassemia caused by body image disturbances experienced so that they feel distrustful and limit their association with their peers, restrictions on activities provided by health workers and parents, and low academic achievement that makes thalassemic adolescents experience psychological and psychological problems, decreased quality of life. Therefore, more attention is needed on the psychological and social welfare of thalassemia adolescents by providing facilities such as counseling and adolescent mentoring programs in order to prevent and reduce psychosocial problems that often occur in adolescents with thalassemia.

References

3. Boonchooduang N, Taylor P Louthrenoo O. Pediatric Hematology and Oncology: Health-Related Quality of Life in Adolescents with Thalassemia Health-Related Quality of Life in Adolescents with Thalassemia. 2015.

