International Journal of Health Sciences and Research
www.ijhsr.org ISSN: 2249-9571

Review Article

Health Related Quality of Life and It's Associated Factors among Thalassemic Children: a Review

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ABSTRACT

Thalassemia is a group of hemoglobinopathies characterized by defect in hemoglobin synthesis. It is chronic disease, where the patients should undergo regular blood transfusion and take iron chelation to maintain their health status. The authors aimed to bring the comprehensive view of overall health related quality of life and find out the various factors associated with their quality of life. The findings from various studies revealed that all the domains of health related quality of life is been affected. The most common factors which had impact on QoL include chelation therapy, repeated hospitalization, educational status of parents and financial factors.

Key words: thalassemia, health related quality of life, factors, chelation therapy

INTRODUCTION

Thalassemia is an autosomal recessive disorder characterized by defect in synthesis of hemoglobin which leads to destruction of red blood cells. [1] The Children with beta thalassemia Major need to undergo repeated blood transfusion throughout their life due to ineffective erythropoiesis. [2] From the detection of first case of thalassemia to the recent advances in therapeutic management, the perception about the diagnostic and treatment modalities for this life debilitating disorder has been drastically changed over the years. But still, whatever intervention, no matter how simple it looks, may alter the level of Health Related Quality of Life (HRQoL) among thalassemic children. Problems associated with disease condition and blood transfusion and other factors results in poor physical and emotional health, disturbed social functions and absent from school or inability to continue schooling. [3]

The level of HRQoL of thalassemic children is influenced by so many factors. There are innumerous studies undertaken throughout the world to assess the impact of thalassemia on the quality of life (qol) among the patients and the factors which promote or hinder the quality of life. The purpose of this review article is to present the overall scenario of health related quality of life and its associated factors among thalassemic children.

METHOD OF REVIEW

Sources of Literature review

In-depth literature review was done using printed sources such as books and journals as well as Google scholar, PubMed and various websites.

Criteria for selection of studies:

The authors used the following criteria for selection of studies for review:

- Studies related to health related quality of life among children with beta thalassemia major
- Studies which have used cross sectional, exploratory research designs (in order to understand the existing condition)
- Comparative studies on quality of life among beta thalassemic children Vs normal counterparts

Total of 17 studies which met the criteria were selected for review. Among which 5 studies were related to assessment of health related quality of life; 4 studies were about comparison of quality of life of thalassemic children with their normal counterparts and remaining 8 studies focused on factors influencing health related quality of life among thalassemic children.

RESULTS

Results of the review are presented under following headings: 1) quality of life of children with beta thalassemia; 2) comparison between QoL of life among thalassemic children and the normal children of same age group and 3) factors associated with quality of life of thalassemic children.

1. quality of life of children with beta thalassemia;

Kaheni S et.al noted that quality of life of children with beta thalassemia major was above average in dimensions such as physical health, psychological health and environmental health. But, qol score was found to be less than average in dimension of social relationship. [4] Similarly, a longitudinal study on qol of βthalassemic children in Chandigarh, India revealed that the most affected domain was social functioning. However the overall QoL found to be improved with progression of illness. [5] But, Ismail M et.al mentioned that the mean total summary scores of QoL in all four dimensions of health were found to be in less thalassemic children and adolescents. They argued that pre-transfusion Hb, frequency of blood transfusion and chelation therapy were the main predictors in HRQoL score among the study participants. [6] In another study, emotional functioning and the school functioning scores were less. School absentism for hospital visits as well as low energy level for performing school activities had a negative impact on the children's HRQoL. [7]

An analytical study on psychological problems and quality of life in thalassemic children revealed that 44% of children had psychological problems and 74% had poor quality of life. This impairment in QoL was found to be due severe difficulties in pain/discomfort dimension (64%), followed by depression and mobility problems of equal severity (33%). [8] Presence of thalassemia as a chronic disorder had negative impact on perceived physical, emotional, social and school functioning of school age children. As per the self report and parent proxy report, emotional functioning domain was the most affected. There was also an association between compliance and quality of life. [9]

2. comparison between QoL of life among thalassemic children and the normal children of same age group

On comparison with the normal siblings, the overall qol is less in thalassemic children and the school functioning is the most affected parameter. [10] Also, the level of qol is found to be less among thalassemic children when compare to QoL of children with other mild illnesses. Majority (80%) of thalassemic children had the feeling that they could not do activities like their friends. [11] When compare to normal healthy children, physical functioning, social functioning and school functioning of thalassemic children was low. The total QoL score was 68.91 in thalassemic children whereas healthy controls had scored 79.79. [12]

A case control survey on assessment of QoL of thalassemia patients revealed that mean physical, emotional, social and school performance as well as overall QoL scores were decreased when compared to the healthy controls. There was a difference

between QoL scores of young thalassemic children and older ones. [13]

3. Factors associated with quality of life of thalassemic children

Thavomcharoensap M et.al in their study on "Factors affecting quality of life in Tai children with thalassemia" identified the following factors as significant which may influence quality of thalassemic children. They are age of child during onset of disease, age at first transfusion, pre-transfusion hemoglobin level, and number of blood transfusions during the past three months, severity of disease and iron chelation therapy. [7] But, other factors such as serum ferritin level, frequency of blood transfusion per year, gender of the child are not significantly associated with the overall functioning score of patients. The study findings confirmed the importance of maintaining pre-transfusion hemoglobin level of at least 9-10.5gms/dl in order to improve the overall quality of life among thalassemic patients. In addition, it was recommended to provide special care to children with severe conditions as well as those who are receiving iron chelation therapy through subcutaneous route.

Various other factors which affect the quality of life of thalassemic children include family income, family history of thalassemia. Mainly, children's poor physical functioning was associated with low family income and poor adherence to treatment and follow up. However thalassemic children's educational status increased their school functionality. [14] Factors like age of patient, educational level of patient's parents, serum ferritin level and complications due to iron overload were significantly associated with physical functioning, overall quality of life and quality adjusted life years of children with thalassemia. [15]

Factors such as Information about the disease condition and family history of thalassemia also had a significant association with all domains of quality of life among school age thalassemic children. [16]

It was also noted that delay in starting of iron chelation therapy found to have negative impact on HRQoL of thalassemic children. [17]

A study on "Better understanding of Health Related Quality of life in Thalassemia patients treated by Iron chelation therapy" identified several factors which influences HRQoL of thalassemic children. The availability of oral iron chelation medicine was found to be the first factor which influences HQoL of these children. It was argued that if there is availability of drug, all patients can became more complaint with the chelation therapy. In addition, it was also noted that health education programs enhanced the awareness about the disease condition among thalassemic children and their parents. So that they became more involved in social networks which had positive effect on children's functioning. Hence, the study findings also recommended that thalassemic children should undergo periodic assessment of quality of life. In addition, intervention programmes especially psychological support need to be planned for support of patients and their families. [18]

Dhirar N, Khandekar J, Bachani D, Mahto D conducted a descriptive cross sectional study to assess the quality of life among children suffering with thalassemia Major. The sample consisted of 241children in the age group of 2 to 18 years receiving care at tertiary level children's hospital in Delhi. The study revealed that the overall quality of life score was low among the study participants and emotional domain was the most affected. The findings also found the following factors are correlated with the quality of life scores: 1) thalassemic children who have other co-morbidities were having poor quality of scores when compared with those who were not having any other conditions. Physical health and psychological domains were severely affected by co morbidities. 2) Children receiving oral iron chelatros had higher QoL scores than those who were taking both oral and subcutaneous therapy.

The painful injection significantly affected the psychosocial health. 4) Increased frequency of blood transfusion per month and more frequent visit to the hospital had negative impact on physical, psychosocial and school activities domains of quality of life in these children. 5) Traveling, painful diagnostic and therapeutic procedures, free health care services were the other major factors had profound impact on overall quality of life among this group. [19]

The frequent visit to health care facility was one of the main factors for poor qol score in school functioning of children with thalassemia major. Also, qol of thalassemic children was substantially influenced by many other factors such as impact of having chronic illness, body image disturbance, subcutaneous iron chelation therapy, complications and importantly uncertainty about their future. [20]

CONCLUSION

The literature reviews related to health related quality of life and associated factors shows that overall quality of life among children with beta thalassemia major was found to be less. However there are variations in type of domain/ dimension affected from one study to another. Also there were so many factors found to have impact on quality of life like frequent hospital visits, therapeutic procedures, socio-economic burden etc. but there was a contradictory between the factors identified in above studies. However, frequent hospitalization and chelation therapy were common factors which influence QoL of thalassemic patients in majority of the studies.

The variation in quality of life scores and influencing factors in various studies may be due to different types of tools and techniques and analytical procedures used by different researchers apart from study participants' demographic characteristics.

REFERENCES

1. Hockenberry M.J, Wilson D. Wong's Essentials of Pediatric Nursing. 8th edition, New Delhi: Elsevier India Pvt Ltd; 2011; 923-4
2. Lakshmanaswamy A. Clinical Pediatrics history taking and case discussion. 4th edition, Haryana: Wolters Kluwer; 2017; 656-62
3. Khani H, Majdi M, Azadmarzabadi E, Montazeri A, Ghorbani A, et al. Quality of life in Iranian Beta-thalassemia major patients of Southern coastwise of the Caspian Sea. Behavioral Sciences. 2009; 2: 325-332.
4. Kaheni S, Yaghobian M, Sharefzadah GH, Vahidi A, Ghorbani H, et al. Quality of Life in Children with B-Thalassemia Major at Center for Special Diseases. Iran J Ped Hematol Oncol. 2013; 3: 108-113.
5. Khaikar P, Malhotra S, Marwaha R. Growing up with families of β thalassemia major using an accelerated longitudinal design. Indian Journal of Med Res. 2010; Oct: 428-37
6. Ismail M, Chun C.Y, Shahar S, Abudul Manaf Z, Rajikan R, Mohd Yusoff N.A, Jamal A.R. Quality of life among thalasemia children, adolescent and their care givers. Sais Malaysiana. 2013; 42 (3): 373-80.
7. Thavorncharoensap M, Torcharus K, Nuchprayoon I, Riewpaiboon A, Indraratna K.. Factors affecting health related quality of life in Thai children with Thalassemia. BMC Blood Disord. 2010; 10: 1-10
8. Shaligram D, Girimaji S.C, Chaturvedi S.K. Quality of life issues in caregivers of Youngsters with thalassemia. Indian J Pediatrics. 2007 Mar; 74 (3): 275-8
9. Dakhakhny M.A, Hesham A.M, Mohamed E.S, Mohamed N.F. Quality of life of school age Thalassemic children at Zagazig city. J of American Science. 2012; 7 (1):186-97
10. Wahyuni S.M, Ali M, Rosdiana N, Lubis B. Quality of life assessment of children with Thalassemia. Pediatrica Indonesia. 2011 May; 51 (3): 163-68.
11. Mutsher. Assessment of health related quality of life upon school age children (6-12) with Thalassemia. Thi-Qar Medical Journal, August 5 (2): 100-108
12. Ismail A, Cambell M.J, Ibrahim M.H, Jone G.L. Health related quality of life in Malasian children with Thalassemia. 2006 4(39): 1-8

13. Hakeem A.G.L, Mousa S.O, Moustafa A N, Mahgoob M.H, Hassan E.E. Health-related quality of life in pediatric and adolescent patients with transfusion dependent β-thalassemiain Upper Egypt (single center study). Health and quality of life outcomes. 2018;16(59): 1-9.
14. Ayoub M.D, Radi S.A, Azab A.M, Abulaban A.A, Balkhovor A.H, Bedair S.W, Ajaouni S.K, Kari J.. Quality of life among children with beta-thalassemia major treated in Western Saudi Arabia. Saudi Med J. 2013Dec; 34 (12): 1281-6.
15. Dahlui M, Hishamshah M.I, Rahman A.J.A, Aljunid S.M. Quality of life in transfusion dependent thalassemia patients on desferrioxamine treatment. Singapore Med J. 2009; 50 (8): 794-9
16. El-Kamah G, Adam S, Magdy P, Michel M, Afifi H. Quality of life outcomes among Egyptian Children with beta-thalassemia. Blood. 2014; 124: 4864. Retrieved from http://www.bloodjournal.org/content/124/21 4864
17. Caocci G, Efficace F, Ciotti F, Ron carolo M.G, Vacca A, PirasE, Littera R et.al. Health related quality of life in Middle Eastern children with beta-thalassemia. BMC Blood disorders. 2012 12:6. Retrieved from http://www.biomedcentral.com/1471-2326/12/6
18. Mahmoud M.A.M. Better understanding of health related quality of life in thalassemia patients treated by iron chelation therapy in the United Arab Emirates. 2015; Retrieved from https://www.duo.uio.no/bitstream/handle/10852/45193/MADIAN-THESIS--FINAL.pdf?sequence=1
19. Dhirar N, Khandekar J, Bachani D, Mahto D. Thalassemia Major: how do we improve quality of life? Springerplus. 2016; Oct 28; 5(1):1895 retrieved from https://www.ncbi.nlm.nih.gov/pubmed/27843752
20. Siddiqui S.H, Ishtiaq R, Sajid F, Sajid R. Quality of life in patients with thas-lassemia major in a developing country. J Coll Physicians Surg Pak. 2014 Jul; 24 (7): 477-80 retrieved from https://www.ncbi.nlm.nih.gov/pubmed/25052969

How to cite this article: Kavitha K, Padmaja A. Health related quality of life and its associated factors among thalassemic children: A Review. Int J Health Sci Res. 2019; 9(5):386-390.

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