

Health Related Quality of Life among Adolescents with Beta-Thalassemia Major: Issues and Concerns

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Abstract:

The present study aims to understand and highlight the issues and concerns of adolescents who are suffering from genetic blood disorder of beta-thalassemia. Beta-thalassemia major is a lifelong disease which can be managed by regular transfusions and iron chelation therapy and includes clinical manifestations like delayed puberty, physical deformations, fragility of bones, fatigue, weakness, to name a few. The physical challenges produce various psychological and social challenges like low self-esteem, social isolation, stress about health and future, depression, hopelessness, etc. This impacts the affected individual's life terribly in many ways. The quality of life being one of them. Health related quality of life is a multidimensional concept which is used to determine well-being in relation to health and disease. In the present study, the health-related quality of life of thalassaemic adolescents between the age range of 14-19 was assessed, and other challenges faced by these patients were discussed in an open-ended interview with them. The quantitative and qualitative analysis of the findings is discussed with respect to the objectives of the study.

Key words: Adolescents, health, quality of life, thalassemia.

Introduction

Being diagnosed with a chronic illness affects all aspects of the individual's life including social relationships, family relationships, activities of daily life, economic well-being and recreational activities. Having a chronic illness makes an individual completely change their lifestyle in order to cope with the manifestations of the condition. Even for an adult this reorientation process can be extremely overwhelming which makes it even more difficult to comprehend what goes on in a child who gets diagnosed with a chronic disease. Beta Thalassemia Major is one such chronic disease. It is the severest sub form of the Beta (β) Thalassemia variant of the Thalassemia group of disorders. It is caused as a result of paucity or faulty synthesis of beta chains causing excess of alpha chains (Schrier, 2002). Also known as Cooley's Anaemia, named after Dr Thomas Benton Cooley (Cooley & Lee, 1925), it can be diagnosed by 6-24 months of age (Brancaleoni, Pierro, Motta, & Cappellini, 2016), with maximum cases of clinical presentation (75-62%) before the age of 1 year (Modell & Berdoukas, 1984). The prime aspect of the disease is the destruction of red blood cells as soon as they are produced by the bone marrow which results in severe anaemia and the only way through which life can be sustained is by regular blood transfusion which continues for the rest of their lives (Nienhuis & Nathan, 2012). The clinical manifestations of the disease include enlargement of spleen and liver that causes a protruding abdomen, pale skin, growth

retardation, slanted mongoloid appearing eyes, poor musculature, depression of the nose bridge, bossing of skull, protruding of gums, thin and brittle bones to name a few (Galanello & Origa, 2010).

As a result, there are a variety of psychological issues faced by the patients throughout their life. In early years, the child does not understand what is happening and feels pain and discomfort of the treatment but he/she develops a trust and confidence in parents or other caretakers. As the child grows and becomes a toddler, he/she wants greater autonomy but soon understands that he/she is restricted in what they can do and more pointedly, what they cannot do that other children his/her age can. Blood transfusions and iron chelation therapy can add to this distress which may manifest in aggressive and rebellious behaviour. Awareness of being different from their peers especially with respect to physical appearance, inability to take part in certain activities that require vitality and physical strength, the curious questions by friends and peers, bullying and absenteeism as a result of regular visits to the hospital for treatment or other health related issues are few of the many problems the child may face when they reach the school going age (Shaligram, Girimaji, & Chaturvedi, 2007). Adolescence is perhaps the most difficult time in the life of an individual with a chronic illness since birth. The idea of spending the rest of their life managing their condition on their own becomes most stark in this age. Apart from the age specific struggles like peer pressures, obsession with appearance, choosing a career, fitting in the social environment and excelling in life, growing liking towards opposite sex, etc., the chronically ill teen also has to deal with health difficulties, adherence to treatment, managing one's disease on their own, to name a few. New challenges are faced in adulthood like making major career decisions, considering higher education, employment, balancing treatment, all which can become particularly exhausting and frustrating to juggle. The prospect of marriage and having a family of their own seems difficult due to the nature of the disease. Lastly, concerns about mortality remain a cause of agony throughout.

Health-Related Quality of Life

The concept of health-related quality of life (HRQOL) was formulated by Schipper (1990) and he defines it as a functional effect of illness and its treatment perceived by the patient. It's a multidimensional construct as it entails three broad domains – Physical, Psychological and Social Functioning that are affected by individual's disease and treatment. It measures the effects of chronic illness, short and long-term disabilities and treatments on individuals. The studies on the HRQOL of β thalassemia major have been scarce in number especially because the advancements in the medical treatment for the disease have only happened in the last 2 decades. It has been seen that frequent hospitalizations for blood transfusion, agonising injections, absence of sexual development, mental disorders, difficulties in social integration, language deficits, memory impairment and the problems of employment decrease the overall quality of life of thalassaemic patients. Beta thalassemia major patients are largely dependent on blood transfusions and iron chelation therapy. These treatment procedures have their own side effects which negatively impact their health. As a result, the frequency of blood transfusions, duration of treatment and being on chelation therapy play an important role in overall HRQOL of these patients. Dhirar, Khandekar, Bachani and Mahto (2016) did a study assessing the HRQOL of beta thalassemia children at Thalassemia Day Care Centre, Delhi, India. Age, presence of comorbidity, fewer visits to the hospital and not being on

iron chelation therapy, were significantly associated with overall HRQOL. Another study on the HRQOL of transfusion dependent thalassemia indicated that the patients scored significantly lower in all domains of HRQOL in comparison to the healthy controls (Cappellini, 2016).

After reviewing the literature, it was observed that the HRQOL is an important construct and is widely used in understanding the related parameters which predict their occurrence, especially with relation to individuals with various chronic illnesses. With respect to beta thalassemia disease, there is scarcity of Indian studies in relation to this construct almost all of them have been on populations other than Indian like in Iran, Maldives, Italy, Bandar Abbas, Egypt, USA, Malaysia, Pakistan, etc. Hence, the present study aims to understand and highlight the issues and concerns of adolescents who are suffering from genetic blood disorder of beta-thalassemia major by assessing and analysing their health-related quality of life in urban Indian society.

Method

Participants: For the present study, a sample of 30 adolescents between the age ranges of 14 to 19 years of age is taken. The sample included 8 females and 32 males. The information is collected from the Thalassaemic Children Welfare Association (Regd.) at Advanced Paediatric Centre at PGIMER, Chandigarh where the adolescent patients are registered and are taking their medical treatment. Adolescents from the age range who have minimum qualification of 10th grade are selected.

Measures:

World Health Organization Quality of Life – Brief Version by WHO (2004) (WHOQOL-BREF) - This scale is the shorter version of WHOQOL-100 scale and has only 26 items. The first two questions are examined separately as they deal with an individual's overall perception of quality of life and perception of their health. The rest of the questions measure HRQOL on the following four domains: physical health, psychological health, social relationships, and environment. The responses to the items are on a 5-point Likert type scale and the responses vary for different items.

Procedure: After receiving permission from Thalassaemic Children Welfare Association at Advanced Paediatric Centre at PGIMER, Chandigarh, the participants were approached and informed of the intent of the study. Individual consent from the participants were taken and they were administered the test one at a time. Based on previous studies, a list of challenges faced by these patients was prepared and an open-ended interview was scheduled with each participant to seek detailed information on their individual concerns related to family, support system, health care, personality, peer group pressures and complexes, body appearance and image, educational, financial etc. the quantitative and qualitative analysis from the obtained data was done for a better understanding of the results.

Results And Discussion

The present study aimed to understand and highlight the issues and concerns of adolescents who are suffering from genetic blood disorder of beta-thalassemia. For this purpose, a sample of 30 adolescents between the age ranges of 14 to 19 years of age were taken. The sample included 8 females and 32 males. The information was collected from the Thalassaemic

Children Welfare Association (Regd.) at Advanced Paediatric Centre at PGIMER, Chandigarh where the adolescent patients were registered and were taking their medical treatment. Adolescents from the age range who have minimum qualification of 10th grade were selected. They were administered the World Health Organization Quality of Life – Brief Version by WHO (2004). Maximum adolescents rated their individual overall perception of quality of life as ‘Good’ with few rating it as ‘Average’ and others rating it as ‘Very Good’. A large number of adolescents were also ‘Satisfied’ with their overall perception of health with few rating it ‘Average’ and one individual rating it as ‘Very Unsatisfied’.

The results indicated that the mean domain score of the sample was 24.9 for Physical Health, 22.4 for Psychological Health, 8.5 for Social Relationships and 30.86 for Environment (Table-I). The mean for the psychological domain was lower than the mean of physical domain. This result is mirrored in the findings of a meta-analytical study by Baraz, Miladinia, & Nouri (2016) on patients with thalassemia where the disease adversely affected the mental component of the quality of life more than the physical component. Another study in Maldives indicated that adolescents with beta thalassemia major had low levels of HRQOL and the mean score for physical domain was higher than mean score for emotional and psychosocial domain (Maheri et al., 2016). Similar results were obtained in another study and it was further indicated that physical, psychological and environment domain of HRQOL produced higher mean scores compared to mean score of social domain (Kaheni, et al., 2013). Prime reason for this is because the social domain has fewer items in comparison to other three domains. Analysis also showed that a large number of adolescents chose not to respond to the sexual activity facet of the social domain and as a result the mean was taken out from the rest of the items of the domain. The Environment domain had the highest mean score. This result echoes the findings of a study by Ali, Tarawah, Al-Hawsawi, Zolaly, & Turkustani (2015) which also indicated a high mean score on this domain among beta thalassemia patients.

The WHOQOL-BREF scale also indicates score on 24 different facets which are grouped under the four main domains of the measure (Table – II). In the Physical health domain, the subjects indicated the following average score on the 7 facets: 3.73 on Activities of daily living, 3.33 on Dependence on medicinal substances and medical aids, 3.5 on Energy and fatigue, 4 on Mobility, 2.8 on Pain and discomfort, 4.03 on Sleep and rest, 3.43 on Work Capacity. In the Psychological health domain, the subjects indicated the following average score on the 6 facets: 3.93 on Bodily image and appearance, 2.3 on Negative feelings, 4.6 on Positive Feelings, 4.03 on Self-Esteem, 3.6 on Spirituality / Religion / Personal beliefs and 3.96 on Thinking, learning, memory and concentration. In the Social domain, the subjects indicated the following average score on the 2 facets: 4.3 on Personal relationships, 4.2 Social Support and 0.8 Sexual Activity (average on only 6 adolescents who chose to answer this item). Lastly, in the Environment domain, the subjects indicated the following average score on the 8 facets: 3.23 on Financial resources, 3.83 on Freedom, physical safety and security, 4.4 on Health and social care: accessibility and quality, 4.33 on Home environment, 3.63 on Opportunities for acquiring new information and skills, 3.76 on Participation in and opportunities for recreation / leisure activities, 3.76 on Physical environment (pollution / noise / traffic / climate), 4.06 on Transport. This result showed that facet Negative feelings had the lowest score followed by Pain and Discomfort, Financial

Resources and Dependence on medicinal substances and medical aids. The highest score was produced by the facet Positive Feelings followed by Health and social care: accessibility and quality, Home Environment, Personal Relationships and Social Support.

The qualitative analysis was also done which included analysing facets that had most unsatisfactory responses in order to highlight the issues and concerns that of the adolescents. Participation in and opportunities for recreation / leisure activities, Financial resources and Negative feelings emerged as the three facets which indicated area on maximum dissatisfaction followed by Freedom, physical safety and security & Spirituality / Religion / Personal beliefs. Other facets like Pain and Discomfort, Energy and Fatigue, Work Capacity, Opportunities for acquiring new information and skills and Physical environment (pollution / noise / traffic / climate) were also indicated by few to be extremely unsatisfactory. Positive Feelings & Health and social care: accessibility and quality were the only two facets which unanimously produced above average level of satisfaction which suggests that the adolescents enjoy their life and are happy with the health care services provided to them at their centre for treatment.

However, the study has a few limitations. The sample size was small and a future study with a much larger sample may provide more clarity. Also, the gender division is not equal and the sample constitutes largely of males. Gender differences in HRQOL of beta thalassemia major adolescents can provide deeper insight and should be incorporated as a factor in future studies. Other socio demographic factors like socio economic status, educational qualification, frequency of blood transfusion, ferritin levels, etc., can also be used to understand their relation with HRQOL in beta thalassemia major adolescents.

Conclusion

The present study highlighted the issues and concerns of adolescents with beta thalassemia major and to understand how the disease affects the HRQOL of these individuals. The results indicated that psychological health domain had lower mean scores compared to physical health domain which suggests that these adolescents have better functioning and are better able to deal with the physical aspects like day-to-day activities, sleep, work, energy, etc. as compared to mental aspects like body image, self-esteem, concentration, emotions, etc. This highlights the need for providing proper psychological assistance to the children so that they can better deal with their struggles as a result of their disease. Despite being able to cope with the physical health aspect, a low score on psychological health suggests that they have an unsatisfactory perception of themselves and their life which can adversely affect their overall quality of life. Highest mean score on environment domain indicates that adolescents are fairly satisfied with their health care facilities and its access and quality, home and locality environment, transport, security, etc. The sample is from Chandigarh and hence the high score on this domain reflects the quality of life experienced in the city. Lastly, qualitative analysis indicated that Participation in and opportunities for recreation / leisure activities, Financial resources and Negative feelings were the major source of worry for these individuals closely followed by Freedom, physical safety, security & Spirituality / Religion / Personal beliefs, Pain and Discomfort, Energy and Fatigue, Work Capacity.

Acknowledgement: The authors acknowledge the Thalassaemic Children Welfare Association (Regd.) at Advanced Paediatric Centre at PGIMER, Chandigarh for their help and support & the sample for their patience and cooperation.

Conflict of Interest: The authors do not have any conflict of interest.

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Table I: Showing the domain wise mean score

Domains	Mean Score
Physical Health	24.9
Psychological Health	22.4
Social Relationships	8.5
Environment	30.86

Table II: Showing the facet wise mean score

Domains		Facets	Mean Score
Physical Health	1	Activities of daily living	3.73
	2	Dependence on medicinal substances and medical aids	3.33
	3	Energy and fatigue	3.5
	4	Mobility	4.0
	5	Pain and discomfort	2.8
	6	Sleep and rest	4.03
	7	Work Capacity	3.43

Psychological Health	1	Bodily image and appearance	3.93
	2	Negative feelings	2.33
	3	Positive feelings	4.6
	4	Self-esteem	4.03
	5	Spirituality/Religion /Personal beliefs	3.6
	6	Thinking, learning, memory and concentration	3.96
Social	1	Personal relationships	4.3
	2	Social support	4.2
	3	Sexual activity	0.8
Environment	1	Financial resources	3.23
	2	Freedom, physical safety and security	3.83
	3	Health and social care: accessibility and quality	4.4
	4	Home environment	4.33
	5	Opportunities for acquiring new information and skills	3.63
	6	Participation in and opportunities for recreation/leisure activities	3.76
	7	Physical environment (pollution/noise/traffic/climate)	3.76
	8	Transport	4.06