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The Compromised Quality of Life in β-Thalassemia Major Children in Non-Urban Setup in a Developing Country

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Abstract

Background: Thalassemia is a chronic autosomal recessive disorder, resulting in ineffective blood formation and chronic hemolytic anemia. Patient of beta thalassemia major is exposed to multiple stresses including multiple transfusions, exposure to hepatitis B and C viruses and multiple psychological and metabolic problems. Assessment was carried out in the form of a survey to find out extent of problems and the measures to improve the quality of life of these patients.

Objectives: To determine the problems faced by thalassemic children and highlight their personal, social and psychological affect so as to assess their quality of life.

Methods and Study Design: A cross sectional study was carried out in Day care Thalassemia center. 50 children aging 7-18 years were enrolled. After recording the subject's demographic and disease details a survey based of SF-36 questionnaire was filled. The correlation with demographic and clinical parameter was evaluated.

Results: A total of 50 patients with the mean age of 15 ± 34 with the distribution of 35 males and 15 females from urban cum rural areas were analyzed. Majority of them were either illiterate or under primary education (92%). All of them were transfusion dependent. Consanguinity of parents was mentioned in 88% of patients. Females secured low scores as compared to males in almost all the parameters. Poor socioeconomic status, non-compliance and presence of multiple complications determined the low scoring in SF-36.

Conclusions: The quality of life in our cohort was found to be compromised and low due to multiple factors such as low family income, bigger families, repeated episodes of illness and anemia. The management can be improved if psychological and financial support is available for the patients.

Keywords: Beta thalassemia major; Quality of life; Multiple transfusions

Introduction

Beta thalassemia major (β-TM) or Cooley's anemia is widely spread over a vast area of Mediterranean countries, India, Pakistan, Iran and whole of Middle East. In Thalassemia Major either both the globin chains of hemoglobin molecule are absent or one the chain is markedly reduced [1]. It's a genetic disease which is characterized by ineffective hemoglobin synthesis, chronic hemolytic episodes, typical facial appearance due to frontal bossing, failure to gain weight and height, repeated transfusion, painful courses of chelation therapy and regular absence from schools [2]. If they don't get access to proper supervised management plan resulting in inadequate and improper management, it is a fatal disease even in very young age [3].

Pakistan has the highest number of transfusion dependent thalassemia children in the whole world [4]. Majority of the population is residing in villages or small towns where the medical facilities are far below the cities. Management of β-TM patients becomes even more difficult. Majority of the patients have access to private clinics where the health staff lacks essential expertise for handling such a demanding disease state. As majority of thalassemia care centers being administered by different NGOs are only in big cities. A large number of patients die at an early stage or receive suboptimal therapy resulting in very poor health [5].

Individuals having exposure to well established thalassemia care centers in cities have dramatic increase in survival and life expectancy due to the introduction of regular blood transfusions and iron chelation regimens [6]. Apart from survival, the quality of life (QOL) from both the social and general health aspect is found to be much lower as compared to the general population [7].

Due to the availability of supervised management plan in urban areas, the life expectancy of β-TM patents has increased considerably with the appearance of magnitude of various problems, not experienced earlier such as variety of psychological disturbances including anxiety, depression and inferiority complex [6]. Others include mal-developed secondary sexual characters, delay in menarche, infertility, multiple complications secondary to the disease process and multiple transfusions, multiple endocrine problems due to deposition of ferritin in the body [8]. Difficulties in completion of education with resulting unemployment, difficulty In finding a life partner with inability to support own family and number of uncertainties of life expectancy [8].

SF-36 is a health survey containing 36 self-explanatory and patient friendly questions for assessing the quality of life in general population as well as in specific groups having chronic illness like β-TM. The reliability of this survey has been approved by multiple investigators. The quality of information based on this survey helped us a lot in finding out many aspects of difficult situation of our patients [9].

Our study aimed to assess the quality of life and social aspects of thalassemia patients which are mostly not acknowledged. Our society should pay special attention to this silent group who is constantly struggling for their health and life. Special programs must be arranged aiming to provide them moral support, professional education and specialist centers all over the country for prenatal diagnosis of such a crippling disorder.

Material and Methods

This cross sectional study was carried out in Thalassemia day care center managed by Arif Memorial teaching hospital in Lahore located in close vicinity of district Kasur, Pakistan. Data of 50 already diagnosed patients of β-TM attending the clinic regularly for blood transfusion and chelation therapy, was collected during the period of June to August, 2015. The patients who were not on regular blood transfusions or having hetrozygosity with other hemoglobins such as C or S or have some chronic disease were regretted for inclusion in the study. Age of the patients ranged from 7 to 18 years.

Modified 36-Item Short Form Survey (SF-36) questionnaire, translated in Urdu was distributed among the patients and their attendant. Permission from the Ethical committee of the hospital for subject study was obtained. All the patients were sought for written consent to participate in the study. Patients were encouraged to fill the questionnaire by themselves but in case of illiteracy or some inability, a trained nurse helped them in recording the answers. Confidentiality of the obtained data was ensured and was used for statistical purposes only.

Statistical analysis was performed using SPSS-10. The significance of association between demographic and clinical data was calculated by Chi square test. Paired t-test was carried out for comparison among certain characteristics. P value <0.05 was considered significant.

Results

A total of 50 patients were enrolled, with the mean age of 15.34 ± 4.2. Eighteen subjects (36%) were aged 10 years or younger and thirty two (64%) of them were older than 10 years of age. 35 subjects were male while 15 were females. Consanguinity of parents was mentioned in 44 (88%) out of 50 patients. All our patients were transfusion dependent. Majority of them (30) required twice monthly transfusions whereas rest, required at least one blood transfusion on monthly basis.

Most of our patents were pursuing primary school education with long periods of absence because of their illness or stay in hospital for transfusions or related complications. 5 of them left schools because of inability to cope with school studies or reluctance on part of the parents. The socio-economic state of 37 (74%) of the families was found to be very low.

Parents of 15 patients mentioned about lack of funds for even transportation to and back from Thalassemia center. NGOs try to provide blood transfusions and iron chelating medication but at times patients have to go back half treated. Other demographic findings are mentioned in Table 1.

Character	Number
Age	
7-10	18
>10	32
Sex	
Male	35
Female	15
Background	
Urban	28
Rural	22
Marital status	
Married	Nil
Single	50
Education	
Primary	46
Beyond	4
Type of living	

Joint	23
With parents	27
Occupation	
Student	32
Employed	1
Unemployed	13
Economic class	
Low	44
Moderate	6
High	
Complication of multiple transfusions	
Severe	8
Moderate	29
Low	13
Blood transfusions	
Regular	28
Irregular	22
Type of chelation	
Desferioxamine	32
Oral	18
Iron chelation therapy	
Good compliance	38
Poor compliance	12

Table 1: Quality of life: Patients characteristics and descriptions.

Descriptive variables	Total (n=50)	Males (n=35)	Females (n=15)	P-value
Physical functioning	82 ± 12.5	87 ± 12.9	76 ± 14.1	0.004
General health	61.2 ± 13/5	67.2 ± 15.5	54.3 ± 15.3	0.006
Socially active	63.4 ± 18.4	72.5 ± 18	54.7 ± 20	0.005
Mental functions	59.3 ± 17.3	63.7 ± 19	55.6 ± 14.9	0.012
Pain episodes	64.5 ± 11	69.6 ± 17	60.3 ± 13.8	0.034
Social relationship	65.9 ± 13.4	73.6 ± 17.7	57.7 ± 15.6	0.002
Vitality	61.5 ± 11.7	66.62 ± 15.5	57.5 ± 15.7	0.005
Total SF-36	65.5 ± 15.1	71.41 ± 13.5	59.7 ± 16.4	0.056

Table 2: Heath related quality of life and its dimensions as evidenced by SF-36 scoring.

The findings mentioned in Table 2 are pertaining to SF-36 scores. As already mentioned majority of our patients could not document their responses by themselves so we provided them help in filling all responses but nobody from the investigation team suggested any answer to any of the patient. Eight scale results include the summary of two status i.e. health pertaining to physical and mental point out view and the total of scores for both the male and female patients of β-TM. No significance was detected between the total scores and physical and mental health status among both the sexes. Body pain and social relationship revealed significantly higher results in male participants.

Discussion

Beta thalassemia major is an incapacitating disease not only life threatening for the patient but drains away the strength and economy of the whole family. It's really painful experience to see the parents of these patients struggling for their loved one knowing that he/she wouldn't be with them for long [10]. The prevalence of consanguinity is very high in Pakistan and according to a recent study in some areas of Punjab and KPK its incidence is on increase. This trend has a definite

role in the high prevalence of thalassemia in Pakistan [11]. In our study 44 patients gave history of consanguinity of their parent marriages. According to a recent study the incidence of consanguineous marriages has been noted up to 64% in certain areas in Pakistan [11]. It is well known fact that all the autosomal recessive disorders including thalassemia and hemoglobinopathies are potentiated by this trend.

The QOL among thalassemia patient is known to be low as compared to the unaffected children of the same age [2]. Such a difference in not an unexpected one as these children have lot of challenges such as special physical and emotional problems related to the disease, repeated visits to hospitals, pathological effects of raised body iron stores resulting in stunted growth, feeble appearance, diabetes, hypothyroid, lack of appearance of secondary sexual characters, infertility and knowingly short life span instead of all possible management measures [8].

Physical functioning was grossly affected in all the β-TM patients more so in the girls. This was probably because many of our patients didn't come regularly for blood transfusions resulting in of severe anemia, repeated episodes infections and febrile illnesses. Most of the families in our cohort were of low socioeconomic status. Quite a number of parents mentioned lack of resources for even transportation of their children to and back from Thalassemia day care center. Many a time blood transfusion needed to be purchased because of non-availability of cross matched blood due to atypical antibodies in the serum of their patient. Similar findings were noted in affected children Seferi et al. [12]. Social activities of the children in our cohort were found to be very low scoring in both the sexes (72.5 and 54.7). All the children were transfusion dependent and about 44% of them with h/o irregular visits with varying degrees of severe anemia and ill health. The studies in developed world revealed much better social activities and well managed treatment schedules with blood transfusions only on weed ends to avoid abstinence form school [13]. In contrast our hospitals are closed on weekends and functional during week days, ensuring repeated absentees from schools. This is probably the reason that out of total fifty, forty six were at the level of primary school and only four of them went beyond that. The feeling of lagging behind other age matched fellows leads to various psychological and social problems including depression, anxiety and isolation etc. [14].

One interesting finding that girls scored low as compared to boys in almost all the assessed parameters such as physical and social activity, general and mental health and vitality surprisingly quite opposite finding was noted by Safizadeh et al. where girls did fairly better in general health domain [7]. Similarly another study by Hadi revealed better QOL in women in multiple SF domains [13]. Studies conducted in Malaysia and Thailand revealed that sex is not an effective determinant on QOL domains [14,15]. The explanation of our finding may be that quite a number of our subjects belong to rural back ground and even children referred as Urban are actually from small towns in close vicinity of Lahore. Traditionally males being the expected earning members of family are preferred over females and boys get more attention as compared to girls.

A large majority of our cohort i.e. 74% presented with moderate to severe complications of repeated blood transfusions. Such as high incidence of Hepatitis B and C leading to liver dysfunction and development of variety of endocrine problems including hypoparthyroidism, hypothyroidism, diabetes mellitus and lack of secondary sexual characters in young boys and girls. Mothers of two of the girls mentioned about the anxiety of their daughter regarding lack of menarche and poorly developed body features. These metabolic and endocrine problems along with repeated anemic state and multiple transfusion lead to growth stunting and markedly dark skin color. This entire scenario leads to further isolation and depression in the thalassemia patients. Similar finding have been observed by multiple investigators where the QOL was affected badly by the isolation and developing psychiatric mind set [16-18].

Another finding was a relatively better QOL in thalassemia receiving oral chelators as compared to those getting injectable [19-21]. Better compliance and painless medication allows better control of Ferritin concentration. Findings of Osborne et al. also claimed better QOL if oral iron chelators were available for thalassemia patients [22].

This is first study in which we tried to focus the problems of lower socio-economic group in both the rural and urban thalassemia children. We need to organize the treatment plan tailored according to patient's need and the efforts in prevention of beta thalassemia major need to be implemented at national level.

Conclusion

The QOL in the surveyed thalassemia major children was found be much below the average children of their age. Multiple factor may be operative in this respect such as low family income, big families, lack of proper treatment, education and physical activities lead to their social and psychological isolation. Situation gets compounded by the development of variety of complications and transfusion related viral infection.

References

1. Kaheni S, Yaghobian M, Sharefzadah GH, Vahidi A, Ghorbani H, et al. (2013) Quality of Life in Children with B-Thalassemia Major at Center for Special Diseases. Iran J Ped Hematol Oncol 3: 108-113.
2. Khani H, Majdi M, Azadmarzabadi E, Montazeri A, Ghorbani A, et al. (2009) Quality of life in Iranian Beta-thalassemia major patients of southern coastwise of the Caspian Sea. Behavioral Sciences 2: 325-332.
3. Telfer P, Constantinidou G, Andreou P, Christou S, Modell B, et al. (2005) Quality of life in thalassemia. Ann N Y Acad Sci 1054: 273-282.
4. Alwan A, Modell B (1997) Community control of genetic and congenital disorders. WHO regional office for eastern Mediterranean, Alexandria, Egypt.
5. Ansari SH, Shamsi TS, Ashraf M, Bohray M, Farzana T, et al. (2011) Molecular epidemiology of β-thalassemia in Pakistan: far reaching implications. Int J Mol Epidemiol Genet 2: 403-408.
6. Verma IC, Saxena R, Kohli S (2011) Past, present and future scenario of thalassaemic care and control in India. Indian J Med Res 134: 507-521.
7. Safizadhe H, Farahmandinia Z, Nejed SS, Pourdamghan N, Araste M (2012) Quality of life with Thalassemia Major and Intermedia in Kerman-Iran. Mediterr J Hematol Infect Dis 4: e2012058.
8. Tahir H, Amna S, Khawaja S, Mahmood T (2011) Complications in Thalassaemia Patients Receiving Blood Tranfusion. J Biomed Sci and Res 3: 339-346.
9. Sobota A, Yamashita R, Xu Y, Trachtenberg F, Kohlbry P, et al. (2011) Quality of Life in Thalassemia: A Comparison of SF-36 Results from the Thalassemia Longitudinal Cohort to Reported Literature and the US Norms. Am J Hematol 86: 92-95.
10. Caro JJ, Ward A, Green TC, Huybrechts K, Arana A, et al. (2002) Impact of thalassemia major on patients and their families. Acta Haematol 107: 150-157.
11. Jehangir K, Arshad A, Bakht TK, Zaheer A, Waqas AS (2015) Impact of Consanguinity on Health in a Highly Endogamous Population in District Buner, Khyber Pakhtunkhwa, Pakistan. J Genet Disor Genet Rep 4: 1-4.

12. Seferi I, Xhetani M, Face M, Burazeri G, Nastas E, et al. (2015) Frequency and specificity of red cell antibodies in thalassemia patients in Albania. Int J Lab Hematol 37: 569-574.
13. Hadi N, Karami D, Montazeri A (2009) Health-related quality of life in major thalassemic patients. Payesh 8: 387-393.
14. Thavorncharoensap M, Torcharus K, Nuchprayoon I, Riewpaiboon A, Indaratna K, et al. (2010) Factors affecting health-related quality of life in Thai children with thalassemia. BMC Blood Disord 10: 1.
15. Ismail A, Campbell MJ, Ibrahim HM, Jones GL (2006) Health related quality of life in malaysian childran with thalassamia. Health and Quality of life Outcomes 4: 39.
16. Osborne RH, De Abreu Lourenço R, Dalton A, Houltram J, Dowton D, et al. (2007) Quality of life related to oral versus subcutaneous iron chelation: a time trade-off study. Value Health 10: 451-456.
17. Elalfya MS, Faridb NM, Labiba JH, RezkAllah HK (2014) Quality of life of Egyptian β-thalassemia major children and adolescents. Egyptian J Hematol 39: 222-226.
18. Cheuk DK, Mok AS, Lee AC, Chiang AK, Ha SY, et al. (2008) Quality of life in patients with transfusion-dependent thalassemia after hematopoietic SCT. Bone Marrow Transplant 42: 319-327.
19. Hongally C, Benakappa DA, Reena S (2012) Study of behavioral problems in multi-transfused thalassemic children. Indian J Psychiatry 54: 333-336.
20. Clemente C, Tsiantis J, Sadowski H, Lee C, Baharaki S, et al. (2002) Psychopathology in children from families with blood disorders: a cross national study. Eur Child Adolesc Psychiatry 11: 151-161.
21. Shaligram D, Girimaji SC, Chaturvedi SK (2007) Psychological Problems and Quality of Life in Children with Thalassemia. Indian J Pediatris 2: 727-730.
22. Stuber ML (1996) Psychiatric Sequelae in seriously ill children and their families. Psychiatr Clin North Am 19: 481-493.