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Student Article

Quality of life in transfusion-dependent thalassemia patients

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الملخص

أهداف البحث: يُعد مرض الثلاسيمية أكثر الاضطرابات الوراثية شيوعا في منطقة البحر الأبيض المتوسط. وعلى الرغم من التطورات الأخيرة في علاج مرض الثلاسيمية، فإن سكان البلدان النامية لا يحصلون على العلاج المناسب. وبالنسبة لمثل هذه الأمراض المزمنة فليس المهم فقط هو البقاء على قيد الحياة، ولكن أيضا جودة الحياة، التي تقودها في المقام الأول القيود النفسية والاجتماعية. تستكشف هذه الدراسة عوامل مختلفة تؤثر على جودة الحياة عند مرضى الثلاسيمية المعتمدين على نقل الدم.

طرق البحث: شملت هذه الدراسة المقارنة، أطفالا يعانون من الثلاسيمية الكبرى ويتلقون نقلا منتظما للدم للسنوات الخمس الماضية. وتمت مطابقة الضوابط بالنسبة للعمر، والجنس، والوضع الاقتصادي الاجتماعي، وكانت الضوابط أطفالا أصحاء فقط. تم تقييم أنواع مختلفة من "جودة الحياة" باستخدام أداة تقييم جودة الحياة لمنظمة الصحة العالمية.

النتائج: شملت دراستنا ٩٠ حالة مصابة (٨ حالات تسرب) و٩٨ من الضوابط (بلا تسرب)، بعمر ٨.٣ ±٤.٤ في الحالات المصابة و١٢.٢±٤.٧ في الضوابط. ومجموع متوسط النتيجة الكلية لجميع الأسئلة بالنسبة للمصابين كان ٨٢.٠٤±١٥.٥٤ بينما كانت ٨٧.٨٦ ±١٢.٩ في الضوابط. وكانت الفوارق بين المصابين والضوابط أكثر وضوحا في الذكور في كل العوامل تقريبا. ولم تكن هناك فوارق ذات قيمة بين المجموعتين بالنسبة لمتغيرات الألم، والمظهر، والعلاقات مع الآخرين.

الاستنتاجات: على الرغم من عدم وجود فرق كبير في درجة جودة الحياة عند أطفال الثلاسيمية، إلا أننا وجدنا فارقا كبيرا بين الذكور مقارنة بالإناث. وتتطلب مؤشرات هذه النتيجة أن تُطرق في دراسات مقارنة أخرى.

الكلمات المفتاحية: أطفال؛ جودة الحياة؛ الثلاسيمية الكبرى؛ عوامل نفسية؛ ألم جسدي

Abstract

Objectives: Thalassemia is the most common genetic disorder in the Mediterranean region. Despite recent advances in the management of thalassemia, people living in developing countries do not receive satisfactory treatment. For such chronic conditions, not only is patients' survival important but their quality of life (QOL) is also important, which is primarily driven by psychological and social constraints. This study explores various factors that affect QOL in transfusion-dependent thalassemia patients.

Methods: This case control study included children with thalassemia major who received regular transfusions for the last five years. Controls were matched for age, gender and socio-economic status and included only healthy children. Different types of QOL were assessed using the World Health Organization (WHO) Quality of Life Assessment tool.

Results: Our study included 90 cases (8 dropouts) and 98 controls (0 dropouts), with an average age of 8.3 ± 4.4 in cases and 12.2 ± 4.7 in the control group. The total mean aggregate score of all patient questions was 82.04 ± 15.54; in the control group, the score was 87.86 ± 12.9. In nearly all factors, differences between cases and controls were most significant in males. There were no significant differences for the variables of physical pain, appearance and relations with others in both groups.

Conclusion: Although there was no significant difference in the QOL score in thalassemia children, a more significant difference was observed in male patents than in females. The implications of this finding must be explored in further case-control studies.

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Keywords: Children; Physical pain; Psychological factors; Quality of life; Thalassemia major

Introduction

Worldwide, thalassemia is a serious public health problem because of the high prevalence extending from the Mediterranean and parts of Africa throughout the Middle East and the Indian sub-continent, Southeast Asia, Melanesia and into the Pacific Islands, ranging from 2% to 25%.[1–3] Each year, 50,000 to 100,000 children die of thalassemia major in low- and middle-income countries, and approximately 7% of the world's population are carriers of a haemoglobin disorder.[4,5]

Despite recent advances in thalassemia management, people living in developing countries do not receive satisfactory treatment.[6] For such chronic conditions, not only patient survival is important but also their quality of life; psychological and social functioning are particular constraints.[7–10] The complications of thalassemia major are known to affect quality of life.[11] According to the WHO definition, quality of life (QOL) is individuals' perceptions of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards and concerns.[12]

Beta thalassemia major is the most severe form of thalassemia, characterized by a severe microcytic, hypochromic iron deficiency.[8] In paediatrics particularly, β-thalassemia major and its complications are associated with noteworthy psychological effects, emotional burdens, hopelessness, and trouble with social integration. Children with thalassemia demonstrate weakened abstract thinking and difficulties with language, consideration, memory, constructional/visual spatial abilities, and executive functions, all of which are more prominent in haemosiderotic subjects.[13,14] These children feel slightly different from their peers and develop negative thinking about life, show more anxiety and possess low self-esteem. Although their behavioural profile is similar to normal people's, a large number of these children demonstrate severe psychological deficits because of trouble following agonizing chelation.[15–18]

Goal

The purpose of the study is to access the quality of life of transfusion-dependent thalassemia patients in the paediatric age group.

Materials and Methods

This case control study selected 98 children (cases) having thalassemia major who were receiving regular transfusions (once every 4–6 weeks minimum) for the last five years; 98 healthy controls from the schools were matched for age, gender and socio-economic status. Written informed consent was obtained from the parents of all participants younger than 18. Quality of life was assessed using the standardised tool WHO-QOL-BRF (Arabic Version) and was conducted in a tertiary care hospital, King Saud Medical City, Riyadh, KSA from March 2016 to February 2017. Ethical approval was granted by the Institutional Review Board under IRB registration number H-01-R-053.

From the total sample of 196 children, we eliminated the records of 8 cases because of inappropriate responses or because participants declined to respond properly after consent was granted. Hence, we had data from 188 records, analysed using SPSS 21.0, and the results are presented as descriptive and inferential statistics using Chi-square and *t*-test for scores of QOL BRF with a 5% level and tested for statistical significance.

Results

The average age in the control group was 12.2 ± 4.7 years and that of the case group was 10.3 ± 4.4 years. We observed that a majority (73.9) of the children had 'good' and 'very

Table 1: Domain 1: Response to physical health domain.

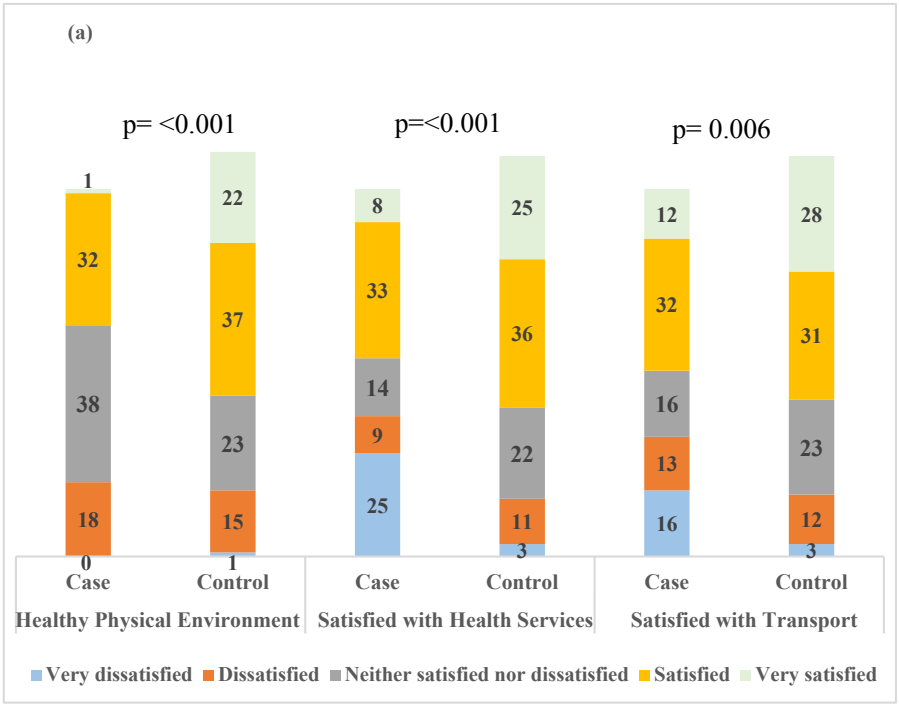
	Status		p-Value	Status		p-Value	Status		p-Value
	Case	Control		Case	Control		Case	Control	
Physical Pain				**Med Treatment**			**Energy Daily Life**		
Not at all	9	44	0.000	3	1	0	3	3	0.765
A little	21	32		23	7		5	7	
A moderate amount	28	16		26	26		27	28	
Very much	21	2		33	38		37	40	
An extreme amount	8	2		4	23		16	20	
General Health				**Satisfied with Activities**			**Able to Get Around**		
Very dissatisfied	3	2	0.000	1	0	0.000	2	5	0.001
Dissatisfied	18	7		13	3		20	7	
Neither satisfied nor dissatisfied	22	16		32	21		21	16	
Satisfied	40	44		35	42		38	40	
Very satisfied	5	28		8	27		8	29	

*Statistically significant at 0.05.

Table 2: Domain 2: Response to psychological health domain.

	Status		p-Value	Status		p-Value	Status		p-Value
	Case	Control		Case	Control		Case	Control	
Satisfied with Sleep				**Satisfied with Capacity**			**Enjoys Life**		
Very dissatisfied	3	2	0.000	7	0	0.009	2	2	0.067
Dissatisfied	18	7		4	5		12	6	
Neither satisfied nor dissatisfied	22	16		13	15		36	40	
Satisfied	40	44		44	42		34	30	
Very satisfied	5	28		19	36		5	19	
Bodily Appearance				**Meaningful Life**			**Able to Concentrate**		
Not at all	14	5	0.004	1	0	0.308	3	0	0.022
A little	26	18		16	15		8	6	
Moderately	28	27		30	27		31	17	
Mostly	17	30		32	32		26	38	
Completely	4	17		10	23		21	35	

*Statistically significant at 0.05.



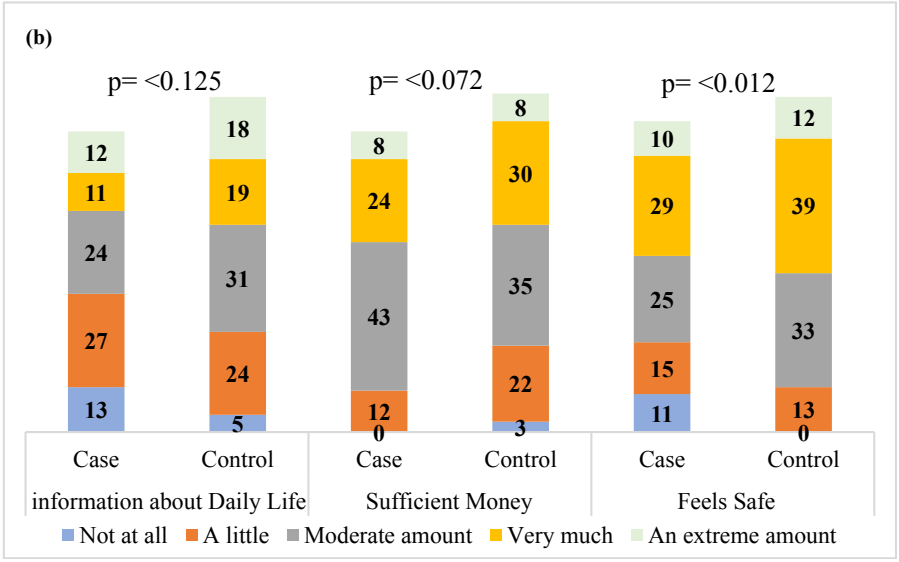


Figure 1: (a) 1 to environmental health domain. (b) Response to environmental health domain.

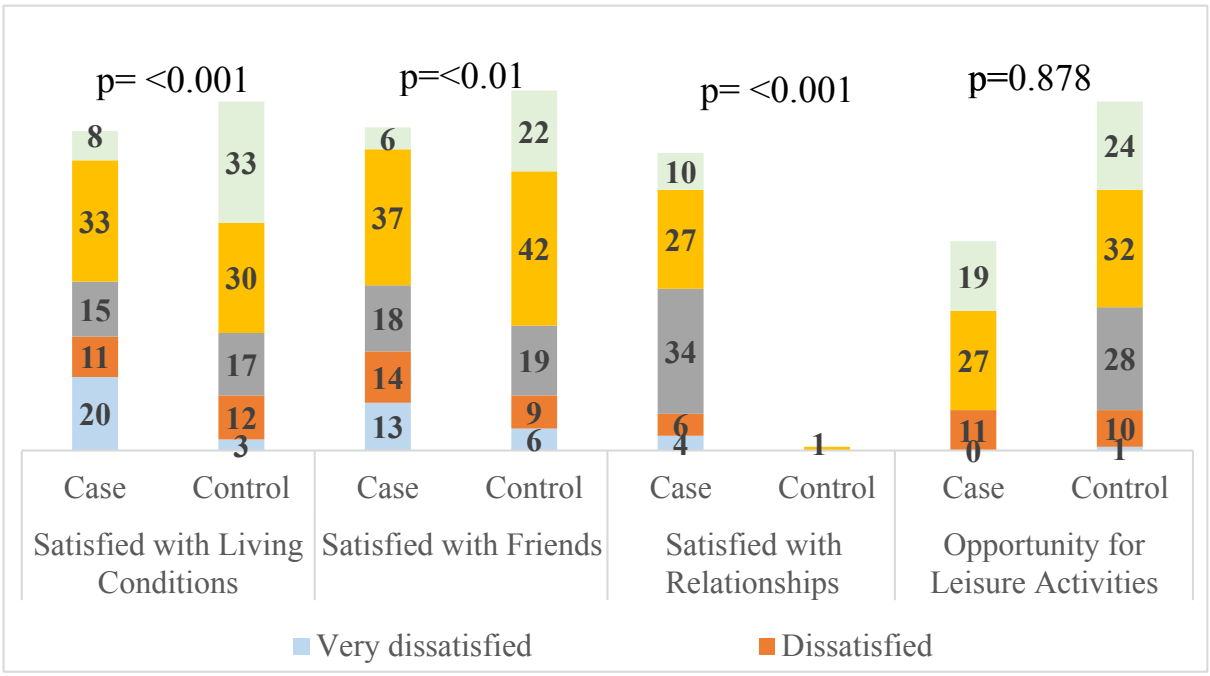


Figure 2: Response to social health domain.

good' QOL with no statistically significant differences between these two groups. However, we observed a dissatisfaction (86.7) with overall general health in both groups.

The total score of all domains was 82.4 ± 5.54 in cases and 87.79 ± 2.91 in controls. Table 1 presents the responses to all determinants of the physical health domain with statistical significance except 'feeling sufficiently energetic to perform daily life activates' among the patients ($p = 0.76$). This result can be attributed to the fact that thalassemia patients have poor oxygen perfusion compared with normal children and thus feel more fatigued and less energetic. Table 2 presents the response to the components of the psychological health domain. We observed that 50 cases believed they led a meaningless life and 55.6 did not enjoy life compared with the controls, with statistical insignificance ($p = 0.30$). Figure 1 presents the distribution of participants for the environmental Health domain. Nearly 48.9 were satisfied with their healthy physical environment, 54.2 with health services and 42.5 felt safe, which was statistically significant ($p = 0.01$).

Figure 2 presents children's satisfaction with Home environment, Health and Social care, Personal relationships and opportunities for recreation and leisure activities. There was a significant association observed among the groups and the components of the social domain. The scores of the various facets in each domain were tested using the student *t*-test. The overall mean score for the physical domain was 24.71 for controls and 23.06 among cases ($p = 0.08$). A low mean score of 18.97 ($p = 0.18$) was observed for the psychological domain and 21.42 ($p = 0.057$) for the environment domain compared to the control group, with no statistical significance. The social domain score of 6.1 was higher among the patients with significance ($P = 0.000$).

Discussion

Various aspects of quality of life, particularly those in the psychological and social domains, are generally ignored. This study probes the effect of frequent transfusions on various aspects of quality of life, including psychological and social aspects.

The WHO-QOL-BRF used in this study showed good validity in terms of content, internal consistency and reliability.[18,19] Although the tool is specified for adults, we used it on a paediatric population. The results indicated no significant difference between the quality of life of normal individuals and those receiving transfusions for thalassemia in both males and females in the physical, psychological and environmental domains. However, significant differences were observed in the social domain. It was surprising to note that the quality of life was reported to be better for children receiving transfusions than for healthy controls. A possible explanation for this could be that children with thalassemia receive more attention, making them feel better socially. Hongally[20] also reported that the patients believed that the disease did not affect their family or social relationships. Ali SS[21] also observed that thalassemia patients had significantly higher scores in the social domain. This finding was not consistent with studies by Pruthi, Naderi, and Ishtiaq.[22–24]

The findings by Behdani et al. and Ansari[25,26] and other reports from KSA varied widely from the results of our study, which reported significant differences in QOL in terms of psychological, social and health-related issues. The encountered differences can be attributed to the selected age groups and the diversity and/or differences in the socioeconomic status of the studied subjects from various countries. Differences in the quality of health care services provided may also contribute to this wide difference. By

contrast, the findings of Kehani et al. were similar to our findings in all domains.[27] This difference can be explained by the age group selected. In a study by Ayoub et al.[28] from western KSA, the quality of life among children with beta thalassemia major was affected by factors such as family income and a family history of thalassemia. Education appeared to increase patient functionality, and supportive measures may improve the quality of life in thalassemic patients.

Strengths and limitations of the study

There are few studies in which cases are compared to controls as they are in our study, particularly among paediatric patients. The majority of the published related studies were descriptive whereas this study used an analytic approach to justify its findings. However, the results are not intended to be generalized because of the limitation of the small sample size and the difficulty in collecting data from children.

Conclusion

It may be said that QOL for patients who must undergo transfusions for thalassemia is not similar to the QOL of the control group in all domains except for the domain of social health, in which the patients appear to have a better quality of life than normal individuals. This finding warrants the need for probing in more detail the aspects of quality of life in thalassemia children using a larger sample size and simpler questionnaire to elicit more factual responses.

Authors' contribution

Idea was conceived by RAA and OMA. Literature review was performed by RAA. MAS ad RAA developed study protocol while data collection which included survey from the participants along with data entry were performed by MA, M (Mauth) KA, M (Mazi) KA and MTA. MA and OMA did data analysis. MKA along with MSA drafted the first version of the paper, which was reviewed, initially by MKA, MSA, then finally MTA, and MKA Final did editing and submission to the journal by RAA.

Conflict of interest

The authors have no conflict of interest to declare.

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