

Factors Affecting Health Related Quality of Life and Its Association with Xmn1 Gene Modifier among Transfusion-Dependent β -Thalassemia and HbE/ β -Thalassemia Adolescent in East Coast Malaysia

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Abstract

Gene modifiers XmnI have significant association with disease severity in HbE/ β -thalassemia patients; however there is no previous study on its association with health related quality of life (HRQoL). The aim of this study is to determine factors affecting HRQoL among transfusion-dependent β -thalassemia and HbE/ β -thalassemia adolescents and its association with Xmn1 gene modifier. A cross-sectional study was conducted among transfusion-dependent β -thalassemia adolescents from government hospitals in East Coast Malaysia. HRQoL scores were measured by using the translated PedsQL 4.0 Generic Core Scale while blood was obtained for genetic modifier analysis. The data were analysed using SPSS version 24.0 and its associated factors were analysed using multivariate regression analysis. A total of 151 patients were recruited. The lowest and highest QOL mean score was school function at 59.69 (16.23) and social function at 77.33 (18.87), respectively. Age of first blood transfusion and Xmn1 gene modifier were significantly associated with physical function ($p=0.022$ and $p=0.008$). Xmn1 gene modifier was also found to be significantly associated with emotional function ($p=0.013$). Frequency of blood transfusion per year was found to be associated with social function ($p=0.002$) while gender and age of first presentation were associated with school function factor ($p=0.009$ and $p=0.009$). There was significant relationship between Xmn1 gene modifiers with physical function and emotional function among transfusion-dependent thalassemia adolescents ($p=0.023$ and $p=0.017$). Xmn1 gene modifiers is significantly associated with HRQoL which affects physical and emotional function among the patients. This study findings also contributed to new development of effective preventive strategies in the future.

Keywords: gene modifiers; thalassemia; psychosocial; quality of life.

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