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PREDICTORS OF HEALTH RELATED QUALITY OF LIFE AMONG CHILDREN AND ADOLESCENTS WITH BETA THALASSEMIA IN THREE HOSPITALS IN MALAYSIA: A CROSS SECTIONAL STUDY

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ABSTRACT

Background: Prognosticating factors associated with health related quality of life (HRQoL) facilitates better treatment outcomes in patients with beta thalassaemia. This study evaluated the predictors of health related quality of life among children and adolescents with beta thalassaemia in Malaysia.

Materials and Methods: A cross sectional study in three public hospitals in Malaysia was conducted between July 2008 and July 2009. All registered patients aged 8 to 18 years with beta thalassaemia and of Malaysian citizenship that attended the haematology daycare centres were invited to participate in this study through their parents. A universal sampling method was used. A pre-tested structured questionnaire was used to record the socio demography, medical information and HRQoL (measured by Paediatric Quality of Life Inventory™ version 4.0). Binary logistic regressions were performed to determine the predictors of HRQoL of patients with beta thalassaemia.

Result: The mean total scale score, physical health summary score and psychosocial health summary score of the participants were 69.76 (SD 13.10), 69.51 (SD 16.64) and 69.93 (SD 14.89), respectively. The psychosocial subscale: the mean emotional functioning, social functioning and school functioning scores were 71.89 (SD 17.47), 78.93 (SD 17.79) and 58.93 (SD 17.93), respectively. The predictors of poor physical health of HRQoL were not on blood transfusion or chelation treatment (adjusted OR=18.29; 95% CI=4.34, 76.86) and presence of side effects from chelation treatment (adjusted OR=9.67; 95% CI=2.15, 43.56). The predictor of poor psychosocial health of HRQoL was duration of thalassaemia less than 10 years (adjusted OR=4.91; 95% CI=1.71, 14.13).

Conclusion: Patients with beta thalassaemia who were not on any blood transfusion or chelation treatment and had side effects from chelation treatment were assoiated with poor physical health of HRQoL, while having thalassaemia less than 10 years was associated with poor psychosocial health of HRQoL.

Keywords: beta thalassaemia, children, adolescent, health related quality of life, Malaysia

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1.0 Introduction

Beta thalassaemia is considered the most prevalent of all human genetic diseases worldwide (Weatherall, 2004). It is an increasing global health problem in the Mediterranean region, Middle East, Indian subcontinent and South East Asia including Malaysia (Vullo, Modell, & Georganda, 1995). As a chronic disease it is associated with regular treatment and complications that increases the burden to the patients, their family and healthcare cost. In Malaysia, it is estimated that between 150 and 350 babies are born with beta thalassaemia each year (Tam, 2005). It is more common among the Malays and Chinese ethnicity.

The chronic impact of the beta thalassaemia and its complications are associated with significant physical and psychosocial ill health. The effects of beta thalassaemia on physical health of children include physical deformity, growth restriction, and delayed puberty. In additon, complications of beta thalassaemia such as cardiac failure and arrhythmia, liver disease and endocrine disorders do impact on the physical health. These would affect not only the children's physical functioning but also the psychosocial health (reprensted by emotional, social and school functionings), which are the domains of health related quality of life (HRQoL) (A. Ismail, Campbell, Ibrahim, & Jones, 2006; M. Ismail et al., 2013; Mikelli & Tsiantis, 2004; Pakbaz et al., 2005; Thavorncharoensap et al., 2010). A study from Malaysia reveals children with thalassaemia had lower HRQoL based on the domains of physical, social and school functioning scores when compared with healthy children (A. Ismail et al., 2006).

Previous studies showed factors that were significantly associated with the lower physical functioning of HRQoL were age at onset of anaemia before 2 years old and age of first transfusion before 4 years old, received blood transfusion in the previous three months prior to assessment, pre-transfusion haemoglobin level of 9 gm/dL or less and on chelation therapy less than 3 times a week (M. Ismail et al., 2013; Thavorncharoensap et al., 2010). Children aged less than 13 years old, pre-transfusion haemoglobin level of 9 gm/dL or less, on monthly blood transfusion and on chelation therapy less than 3 times a week were associated with lower psychosocial health scores of HRQoL(M. Ismail et al., 2013; Thavorncharoensap et al., 2010).

Assessment on the components of psychosocial health of HRQoL showed pre-transfusion haemoglobin level of 9 gm/dL or less was associated with lower emotional functioning scores (Thavorncharoensap et al., 2010). Children aged less than 13 years old, age at onset of anaemia before 2 years old and age of first transfusion before 4 years old, and pre-transfusion haemoglobin level of 9 gm/dL or less were associated with lower social and school functioning scores. Age and patients with severe conditions (defined as patients whose age at onset of anaemia before 2 years old and age of first transfusion before 4 years old, pre-transfusion haemoglobin level of less than 7 gm/dL and diagnosed with homozygous beta thalasaemia) were significant predictors of HRQoL (Thavorncharoensap et al., 2010). Based on our literature search, very few studies assessed the predictors of HRQoL in children and adolescent with beta thalassaemia and none in Malaysia. Therefore, this study aimed to determine the predictors of health related quality of life of children and adolescent with beta thalassaemia in Malaysia.

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2.0 Materials and Methods

A cross sectional study was conducted in three public hospital in Selangor, Malaysia between July 2008 and July 2009. These hospitals were choosen because of the relatively larger number of beta thalassaemia patients among all the hospitals in Selangor. Each hospital has a designated haematolgy daycare centre for thalassaemia patients' follow-up and those requiring regular transfusion. A total of 104 patients with beta thalassaemia aged 8 to 18 years old were registered at these three hospitals. The inclusion criteria for this study were registered patients aged 8 to 18 years, with beta thalassaemia and of Malaysian citizenship who attended the three hospitals haematology daycare centres. Those who had other types of thalassaemia such as alpha thalassaemia, and who were unwell on the day of recruitment were excluded from this study. A universal sampling method was used where all registered patients aged 8 to 18 years with beta thalassaemia who fulfilled the criteria were invited to participate in this study through their parents.

A pre-tested questionnaire was used in this study, which comprised three sections: 1) the socio demography, 2) medical information and 3) Paediatric Quality of Life Inventory^{TM} (PedsQL^{TM}) version 4.0. The questionnaire was back translated from English to Bahasa Melayu and back to English. Two sets of questionnaires for the two age group (8- 12 years and 13-18 years) were used in view of a slight difference in the social functioning domain of PedsQL version 4.0. The socio-demography had six questions, which comprised age, sex (male or female), race (Malay, Chinese, Indian or others), religion (Islam, Buddhist, Hindu, Christian or others) and education level (primary school, secondary school or no formal education). In addition, family's information was also included such as family monthly income, number of siblings, and number of siblings with beta thalassaemia and marital consanguinity, which were obtained from the parents or guardian. The patient's medical information included types of beta thalassaemia, duration of thalassaemia, types of treatment, frequency of blood transfusion and/or chelation treatment (subcutaneous desferrioxamine), side effects of chelation treatment (such as pain at injection site, nausea, vomiting, and rashes), complications related to thalassaemia and presence of other medical problem. These information was obtained from the parents or guardian and was verified using the patient's medical record.

The dependent variable in this study was health realted quality of life measured using the PedsQL^{TM} version 4.0 (Varni, Burwinkle, Seid, & Skarr, 2003). It is a valid and reliable 23-item generic scale to measure health related quality of life (HRQoL) in children and adolescent with acute or chronic health conditions. It comprises developmentally appropriate child self-reported forms (ages ranges 5-7, 8-12, and 13-18 years), and parent proxy-reports (ages ranges 2-4, 5-7, 8-12, and 13-18 years). It assessed the essential core domains for paediatric HRQoL measurement: 1) Physical Functioning (8 items), 2) Emotional Functioning (5 items), 3) Social Functioning (5 items) and 4) School Functioning (5 items), for the past one month. These yield the total score and two summary scores, which are the physical health summary score (comprised the physical functioning) and the psyschosocial health summary score (comprised the emotional functioning, social functioning anf school functioning). The responses of each item are measured using a 5-point Likert scale ranging from 0 (never a problem) to 4 (almost always a problem). The items are reversed scored and linearly transformed to a 0 to 100 scale. Higher scores indicate better HRQoL. If more than 50% of the items in the scale were missing, the scale score should not be computed (Varni et al., 2003). In this study, we used the child self-report form. The Malay version of this scale has been validated and has been found to be sensitive and reliable to be used in the local context

(A. Ismail et al., 2006). The content of questionnaire was reviewed by a paediatrician, a family physician and a public health specialist. The questionnaire was pre-tested among 15 children with alpha thalassaemia to assess the degree of understanding when used among children and adolescent with an overall cronbach alpha of 0.97.

The sample size was calculated using the formula to determine mean from single sample based on the average scores of the four domains of PedsQL scale, total scale score and psychosocial health summary from previous study(A. Ismail et al., 2006) with 80% power and statistical significant level of 5%. The estimated sample size was 70 based on social functioning mean scores of 74.29 (SD18.77) and after considering 30% non-respondents. The data was analyzed using Statistical Package for the Social Sciences (SPSS) version 21.0, with a statistical significant p value set at less than 0.05. The socio-demographic characteristics of the respondents, medical information and the health related quality of life (HRQoL) were described as mean and standard deviation (SD) and as frequency and percentage. The HRQoL of the participants were described as mean and SD based on the total scale, physical health summary, and psychosocial health summary scores and the subscale scores of the psychosocial health summary.

Independent t-tests and One way ANOVA were performed to determine the association between HRQoL (based on all the scores) and the socio-demographic characteristic and medical information. Binary logistic regressions were performed to determine the predictors of HRQoL (represented by the physical health summary and psychosocial health summary scores) of patients with beta thalassaemia. The participants' mean scores of equal or greater than this study's sample physical health summary and psychosocial health summary mean scores were used to determine the better HRQoL in these analyses. The variables from the bivariate analyses were selected using the stepwise forward likelihood ratio with 0.20 signifcant levels for an addition of the variable to predict HRQoL. The results were adjusted for age, sex and race and we presents the adjusted odds ratio, 95% confidence intervals and p values.

This study was registered with the Malaysian National Medical Research Registry, Ministry of Health, Malaysia and the Medical Research Ethics Committee, Ministry of Health, Malaysia (NMRR-08-1446-2911) and Universiti Putra Malaysia Medical Ethics Committee (MJKEtikaPer/F01-Lect_Sept08(06)) approved the methods and materials constructed for this study. Informed consents were obtained from patients and their parents.

3.0 Result

3.1 Response rate

A total of 90 patients attended the daycare centres of the three hospitals during the study and 72 were eligible to participate in the study. 70 agreed to participate in this study with a response rate of 97.2%.

3.2 Characteristics of participants

Table 1 summarises the characteristics of this study participants. Most of the participants aged 8-12 years (72.9%) with the mean age was 11.00 (SD 3.33) years and 52.9% were male participants. Most of the participants were of Malay ethnicity (87.1%) and were studying in primary school (72.9%). The average gross monthly family income was RM 2152.14 (SD1126.51). The proportion of participants with beta thalassaemia major, beta thalassaemia

intermedia, HbE beta thalassaemia and beta thalassaemia minor were 58.6%, 22.9%, 10.0% and 8.6%, respectively. The duration of beta thalassaemia was 10.76 (3.36) with 41.4% of the participants had the illness for 10 years and more. A total of 64 participants (91.4%) were receiving blood transfusion with 68.8% received 6 or more transfusion in the last year and 70.0% were on chelation treatment. Of those on chelation treatment, 83.7% were on 5 or more times in a week. 24.5% of the participants experienced side effects from the chelation treatment and 32.9% had some complications related with beta thalassaemia. These included iron overload (11.4%), splenomegaly (10.0%), both iron overload and splenomegaly (8.6%), hepatosplenomegaly (1.4%) and diabete mellitus (1.4%) (not shown in table).

Table 1: Participants characteristics

Variables	
Age (n=70)	
8 – 12	51 (72.9)
13 – 18	19 (27.1)
Sex (n=70)	
Male	37 (52.9)
Female	33 (47.1)
Race (n=70)	
Malay	61 (87.1)
Chinese	4 (5.7)
Indian	3 (4.3)
Others	2 (2.9)
Education level (n=70)	
Primary school	51 (72.9)
Secondary school	19 (27.1)
Mean monthly household income (SD), RM (n=70)	2152.14 (1126.51)
Number of siblings with beta thalassaemia (n=70)	
None	46 (65.7)
≥ one person	24 (34.3)
Marital consanguinity (n=70)	
Yes	7 (9.5)
No	63 (90.5)
Type of β thalassaemia (n=70)	
β thalassaemia major	41 (58.6)
β thalassaemia intermedia	16 (22.9)
β thalassaemia minor	6 (8.6)
HbE β thalassaemia	7 (10.0)
Duration of illness (n=70)	
< 10 years	41 (58.6)
≥ 10 years	29 (41.4)
Types of treatment (n=70)	
Blood transfusion	14 (20.0)
Blood transfusion & chelation treatment	50 (71.4)
Not on any treatment	6 (8.6)
Frequency of blood transfusion/year (n=64)	
< 6 times	20 (31.3)
≥ 6 times	44 (68.8)

Frequency of chelation treatment/week (n=49)	
< 5 times	8 (16.3)
≥ 5 times	41 (83.7)
Side effects of desferrioxamine treatment (n=49)	
Yes	12 (24.5)
No	37 (75.5)
Types of side effects from desferrioxamine treatment (n=12)	
Pain at injection site	7 (58.3)
Nausea and vomiting	5 (41.7)

Data are mean (SD) or frequency (%)

3.3 Health related quality of life

The HRQoL self-report scores for the participants are presented in Table 2. The mean total scale score, physical health summary score and psychosocial health summary score of the participants were 69.76 (SD 13.10), 69.51 (SD 16.64) and 69.93 (SD 14.89), respectively. The psychosocial subscale revealed the mean emotional functioning, social functioning and school functioning scores were 71.89 (SD 17.47), 78.93 (SD 17.79) and 58.93 (SD 17.93), respectively.

Table 2: Health related quality of life scores

Scale	Mean (SD)
Total scale score	69.8 (13.1)
Physical health summary score	69.5 (16.6)
Psychosocial health summary score	69.9 (14.9)
Emotional functioning	71.9 (17.5)
Social functioning	78.9 (17.8)
School functioning	58.9 (17.9)

3.2.1 Factors associated with health related quality of life

Tables 3 presents the HRQoL scores based on the sociodemography and medical information of the participants. There were significant associations between total scale score and age (p= 0.042), race (p= 0.030), education level (p= 0.042), duration of thalassaemia (p=0.006), types of treatment (p<0.001) and presence of side effects from chelation treatment (p=0.006). The physical health summary score was significantly associated with types of treatment (p=0.001) and presence of side effects from chelation treatment (p=0.001). Significant associations between psychosocial health summary score and age (p=0.037), education level (p=0.037), duration of thalassaemia (p=0.003) and types of treatment (p=0.005).

Based of the psychosocial health summary subscales, emotional functioning was significantly associated with age (p=0.003), education level (p=0.003), duration of thalasaaemia (p=0.003) and types of treatment (p=0.002). There was a significant association between social functioning and age (p=0.014), race (p=0.011), education level (p=0.014), duration of thalassaemia (p=0.001) and types of treatment (p=0.017). School functioning was only associated with frequency of blood transfusion (p=0.039).

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Table 3: Health related quality of life according to demograhic and medical information

Characteristics	Quality of life scale					
	Total scale score	**Physical health summary score**	**Psychosocial health summary score**	**Emotional functioning**	**Social functioning**	**School functioning**
Age (n=70)						
8 – 12	67.8 (13.4)	68.1 (16.9)	67.7 (14.6)	68.1 (16.5)	75.8 (18.1)	59.1 (17.9)
13 – 18	74.9 (11.0)	73.2 (15.6)	75.9 (14.4)	81.8 (16.4)	87.4 (14.2)	58.4 (18.5)
p-value	0.042*	0.262	0.037*	0.003*	0.014*	0.886
Sex (n=70)						
Male	69.5 (14.1)	69.4 (18.1)	69.5 (14.8)	72.4 (17.1)	78.5 (17.9)	57.6 (18.2)
Female	70.1 (12.1)	69.6 (15.1)	70.4 (15.2)	71.2 (18.1)	79.4 (17.9)	60.5 (17.8)
p-value	0.845	0.965	0.801	0.773	0.838	0.505
Race (n=70)						
Malay	70.8 (11.9)	69.9 (16.3)	71.3 (13.7)	72.7 (16.6)	80.8 (15.9)	60.4 (17.2)
Chinese	73.9 (13.9)	82.0 (7.4)	69.6 (20.5)	75.0 (21.9)	81.3 (23.9)	52.5 (25.9)
Indian	51.0 (21.9)	47.9 (20.3)	52.8 (23.6)	65.0 (30.4)	51.7 (20.8)	41.7 (20.8)
Others	57.6 (13.8)	65.6 (13.3)	53.3 (14.1)	50.0 (20.4)	57.5 (24.8)	52.5 (17.7)
p-value	0.030*	0.055	0.490	0.280	0.011*	0.268
Education level (n=70)						
Primary school	67.8 (13.4)	68.14±16.94	67.7 (14.6)	68.1 (16.5)	75.8 (18.1)	59.1 (17.9)
Secondary school	67.8 (11.0)	73.19±15.64	75.9 (14.4)	81.8 (16.4)	87.4 (14.2)	58.4 (18.5)
p-value	0.042*	0.262	0.037*	0.003*	0.014*	0.886
Monthly household income (n=70)	69.6 (13.3)	69.3 (10.5)	69.7 (15.3)	71.7 (22.1)	75.0 (20.3)	62.5 (19.7)
< RM 2000.00	69.8 (13.2)	69.5 (17.2)	69.9 (14.9)	71.9 (17.2)	79.3 (17.7)	58.6 (17.9)
≥ RM 2000.00	0.969	0.971	0.971	0.978	0.575	0.614
p-value						
Number of siblings with beta thalassaemia (n=70)						
None	68.9 (14.3)	68.1 (17.2)	69.3 (16.9)	73.4 (17.9)	76.2 (19.3)	58.3 (20.5)
≥ one person	71.5 (10.6)	72.1 (15.6)	71.1 (10.4)	68.9 (16.7)	84.2 (13.3)	60.2 (11.8)
p-value	0.436	0.344	0.636	0.320	0.075	0.670

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Characteristics	Quality of life scale					
	Total scale score	Physical health summary score	Psychosocial health summary score	Emotional functioning	Social functioning	School functioning
Marital consanguinity (n=70)						
Yes	70.0 (6.9)	66.5 (11.1)	71.9 (6.8)	71.4 (14.9)	85.7 (12.4)	58.6 (14.1)
No	69.7 (13.6)	69.8 (17.2)	69.7 (15.56)	71.9 (17.8)	78.2 (18.2)	58.9 (18.4)
p-value	0.955	0.620	0.715	0.947	0.291	0.956
Duration of illness (n=70)						
< 10 years	66.2 (13.8)	67.4 (17.9)	65.6 (14.9)	66.8 (16.6)	73.1 (18.4)	56.8 (17.9)
≥ 10 years	74.8 (10.3)	72.5 (14.4)	76.1 (12.7)	78.9 (16.4)	87.2 (13.1)	61.9 (17.8)
p-value	0.006*	0.205	0.003*	0.003*	0.001*	0.247
Types of treatment (n=70)						
Blood transfusion only/with chelation treatment	71.4 (12.0)	71.5 (15.3)	71.4 (14.1)	73.8 (16.6)	80.5 (16.9)	59.9 (17.2)
Not on any treatment	51.9 (11.3)	48.4 (16.7)	53.9 (15.1)	50.8 (13.2)	62.5 (20.9)	48.3 (23.4)
p-value	<0.001*	0.001*	0.005*	0.002*	0.017*	0.131
Frequency of blood transfusion/year (n=64)						
< 6 times	74.8 (12.2)	72.0 (18.9)	76.4 (13.6)	79.0 (16.1)	83.3 (16.5)	66.5 (14.7)
≥ 6 times	69.9 (11.8)	71.2 (13.6)	69.2 (13.9)	71.5 (16.4)	79.2 (17.1)	56.9 (17.6)
p-value	0.134	0.849	0.059	0.092	0.378	0.039*
Frequency of chelation treatment/week (n=49)						
< 5 times	77.4 (9.1)	78.9 (8.6)	76.7 (10.1)	76.9 (18.5)	87.5 (12.5)	65.6 (11.5)
≥ 5 times	70.8 (11.7)	73.0 (13.5)	69.6 (13.6)	72.2 (15.9)	80.1 (16.1)	56.6 (18.0)
p-value	0.138	0.243	0.173	0.461	0.227	0.181
Side effects of chelation treatment (n=49)						
Yes	64.1 (12.2)	63.5 (10.5)	64.4 (14.7)	68.3 (16.8)	75.4 (20.4)	49.6 (14.5)
No	74.4 (10.2)	77.4 (11.9)	72.8 (12.3)	74.5 (15.9)	83.2 (13.6)	60.8 (17.5)
p-value	0.006*	0.001*	0.056	0.260	0.134	0.051

Peds QL version 4.0; *p-value <0.05

3.2.2 Predictors of health related quality of life

The logistic regression analyses on the physical health of HRQoL and psychosocial health of HRQoL are presented in Table 4 and 5. Not on blood transfusion or chelation treatment (adjusted OR=18.29; 95% CI=4.34, 76.86) and presence of side effects from chelation treatment (adjusted OR=9.67; 95% CI=2.15, 43.56) were predictors of lower physical health summary score. Predictor of lower psychosocial health summary score was duration of thalassaemia less than 10 years (adjusted OR=4.91; 95% CI=1.71, 14.13).

Table 4: Logistic regression analysis on predictors of physical health of HRQoL

Predictors	Beta	SE	Adjusted OR	95% CI	p-value
Not on blood transfusion or chelation treatment	2.90	0.73	18.29	4.34, 76.86	<0.001*
Have side effects of chelation treatment	2.27	0.77	9.67	2.15, 43.56	0.003*
Constant	-1.17	0.38	0.31		

Nagelkerke R^2 = 0.40; *p<0.05, OR=Odds ratio, 95% CI= Confidence interval. The results were adjusted for age, sex and race.

Table 5: Logistic regression analysis on predictors of psychosocial health of HRQoL

Predictors	Beta	SE	Adjusted OR	95% CI	p-value
Duration of beta thalassaemia less than 10 years	1.59	0.54	4.91	1.71, 14.13	0.003*
Constant	-1.15	0.43	0.32		

Nagelkerke R^2 = 0.17; *p<0.05, OR=Odds ratio, 95% CI= Confidence interval. The results were adjusted for age, sex and race.

4.0 Discussion

Our study showed that the total scale, physical health summary and psychosocial health summary scores were similar but the subscale of the psychosocial health summary score revealed school functioning score was the lowest. These HRQoL scores were similar to those found by other studies in Malaysia (A. Ismail et al., 2006; M. Ismail et al., 2013), which average scores approximately 69.0. However, these scores were lower when compared to the study in Thailand (average scores between 75.0 and 78.0) (Thavorncharoensap et al., 2010). Most of the participants in our study and the previous studies from Malaysia were homozygous beta thalassaemia and transfusion dependent as compared to the participants in the Thailand study and this could explain the observed differences in the scores of HRQoL domains.

This study found those who were not on blood transfusion or chelation treatment was a predictor of poor physical health of HRQoL in contrast to a study in Thailand (Thavorncharoensap et al., 2010), where they found children who receive blood transfusion had lower physical health summary scores. The physical health summary score is based on the physical functioning, which assessed the level of physical activity and energy level of the participant's over past month. Most of the patients in our study are transfusion dependent and they would experience symptoms of anaemia such as fatigue and weakness few days before their presentation to their clinic's follow-up for the frequent blood transfusion. Hence, this might affect negatively their physical function. However, in our study the proportion of

patients not on any treatment was small (less than 10%), which could have overestimated the predictive effect on the physical health of HRQoL.
We found that presence of side effects from chelation treatment was a predictor of physical health HRQoL. Similarly, previous studies also found that the burden of the subcutaneous injections of iron chelation treatment 5 to 7 days a week was associated with impaired HRQoL (Shaligram, Girimaji, & Chaturvedi, 2007; Thavorncharoensap et al., 2010). In addition, it is known that among the common side effects of chelation treatment are pain at the injection site, nausea and vomiting (Dubey, Parakh, & Dublish, 2008). This could have contributed to the reduced scores in our study as most of our patients received five or more times in a week of chelation treatment.

Duration of thalassaemia of less than 10 years was a predictor of poor psychosocial health of HRQoL in our study. No previous study assessed duration of illness as a predictor of HRQoL to allow us to make a comparison. In the present study, those with duration of thalassaemia of less than 10 years were those aged less than 13 years old. The younger patients would have less experience in dealing with their illness and may have difficulty understanding the disease they have, which could affect them emotionally. Furthermore, in our study more of the children aged less than 13 years (68%) were on blood transfusion than the adolescents (32%). Therefore, their social and school functions which could also be affected as they need to be away from school to receive the transfusion. Hence this could affct their psychosocial health. The older children could have more knowledge about the disease or were able to understand the support from their parents, carers or peers to allow them to cope better. It has been shown that illness knowledge and social support were positively correlate with self-care behavior in patients with beta thalassaemia major in Taiwan (Yang, Chen, Mao, & Lin, 2001). However, in our study we did not measure social support and knowledge on thalassaemia, which could be the mediators to HRQoL.

There are several limitations to this study. The sample was small and may have caused overestimation of the odds ratio on predicting the HRQoL in children with beta thalassaemia , hence, the results should be interpreted with caution. In addition, the sample is not representative of children and adolescent with beta thalassaemia in Selangor or Malaysia. Further, there were some variables, which may be mediators of HRQoL such knowledge of beta thalassaemia, psychological conditions and social support were not measured in this study, which could explain other possible predictors of HRQoL.

5.0 Conclusion and recommendation

In conclusion, this study identified the predictors of HRQoL in children and adolescent with beta thalassaemia. The predictors for physical health of HRQoL were not on any blood transfusion or chelation treatment and presence of side effects of chelation treatment, while duration of thalassaemia less than 10 years was the predictor for psychosocial health of HRQoL. A close contact with the patients and family/carers through a multi-discipline team approach that involves the health care providers and school would enhance the HRQoL and improve health outcomes of children and adolescent with beta thalassaemia. In addition, future studies are to also evaluate other mediators of HRQoL such psychosocial condition and social support, to allow a comprehensive measure of health related quality of life.

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Declaration

Author(s) declare that

i. The article mentioned above has not been published or submitted for publication in any other journal;
ii. We also declare that the authorship of this article will *not* be contested by anyone whose name is not listed here;
iii. We declare that we contributed significantly towards the research study *i.e.*, conception, design, analysis and interpretation of data and to (b) drafting of the article or revising it critically for important intellectual content;
iv. There is no conflict of interest on this article.

Authors contribution

The first and third authors were responsible for the conception and design of the study. The second author compiled the data and together with the first author analysed and interpreted the data. The first author prepared the first draft of the manuscript. All authors contributed to the subsequent drafts and approved the final version of the manuscript.

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