FACTORS AFFECTING HEALTH-RELATED QUALITY OF LIFE AMONG PAEDIATRIC PATIENTS WITH THALASSEMIASIA: A REVIEW OF LITERATURE

Wan Adnan Wan-Nor-Asyikeen¹, Ab Hamid Siti-Azrin¹, Maryam Mohd Zulkifli², Bin Alwi Zilfalil³

1. Unit of Biostatistics and Research Methodology, School of Medical Sciences, Universiti Sains Malaysia, 16150, Kubang Kerian, Kelantan, Malaysia.
2. Department of Family Medicine, School of Medical Sciences, Universiti Sains Malaysia, 16150, Kubang Kerian, Kelantan, Malaysia.
3. Department of Paediatrics, School of Medical Sciences, Universiti Sains Malaysia, 16150, Kubang Kerian, Kelantan, Malaysia.

Abstract

Thalassaemia is a hereditary blood disorder that is becoming a major health problem all over the world. This chronic illness harms the quality of life of the sufferers by interrupting their physical activities, school performance and social life. Hence, this review takes aim to assess the factors affecting the quality of life of thalassaemia among paediatrics patients. A comprehensive electronic search was conducted by using PubMed, Google Scholar and Science Direct. The search was limited to those articles written in English language and by using Pediatrics Quality of Life Inventory (PedsQL™) 4.0 generic core scale questionnaire only. This review notifies emerging knowledge regarding the factors affecting the quality of life among thalassaemia patients and its implications in the essential core domains for paediatrics health-related quality of life measurements: physical, emotional, social and school functioning. It also empowers a better understanding regarding thalassaemia and assists as a foundation for the development of the effective preventive strategies for it.

Keywords: Thalassaemia, Paediatrics, Factors, Quality of life, Review

Corresponding Author: Dr. Siti Azrin Ab Hamid, Unit of Biostatistics and Research Methodology, School of Medical Sciences, Universiti Sains Malaysia, 16150, Kubang Kerian, Kelantan, Malaysia
Tel: +609-7676832
Email: ctazrin@usm.my

Introduction

Thalassaemia is a genetic blood disorder where it is portrayed as incomplete or no production of one or more globin chains [1]. It is an acute life-limiting, and potentially life-threatening health problem covered all over the Mediterranean zone, the Middle East, the Indian subcontinent and Asian origins as well as in Malaysia [2].

© 2017 MJPCH. All Rights Reserved.
In Malaysia itself, thalassaemia can be seen mainly among Malay and Chinese population [3]. Almost 4.5% of Malaysians population are beta-thalassaemia carriers [4] and up to 40% are HbE carriers [5]. Malaysian Thalassaemia Registry as of September 2011 indicated a total of 5,115 registered thalassaemia patients and of these, 2,207 have beta-thalassaemia major, and 1,594 have HbE-beta thalassaemia [6]. The Ministry of Health of Malaysia expected that each year, between 150 to 350 babies are born with thalassaemia [7].

Malaysian thalassaemia patients have free access to blood transfusions and chelation agents in the form of subcutaneous deferoxamine and oral deferiprone in government hospitals all over the country. Starting from the year 2012, the newer oral chelator, deferasirox was accessible especially for the younger patients. However, most of the patients still obtained the blood transfusions and iron chelation in the form of subcutaneous deferoxamine.

Children with thalassaemia appear well at birth. However, due to the incomplete or absence of adult haemoglobin, anaemia is tended to develop and then becomes progressively worse. If thalassaemia patients do not receive an appropriate treatment, it can end with an early death [8]. On the other hand, for those children that do survive, this problem has serious implications towards their health-related quality of life (HRQoL) and may disturb their education and social activities. Children with thalassaemia need to go through blood transfusions and they also have to obtain desferal injections for iron chelation therapy to eradicate excessive iron in the blood, which resulted from the blood transfusions [8, 9].

The impact of beta-thalassaemia and its serious complications linked with physical and psychosocial health problems. Beta-thalassaemia gives an effect on the children’s physical health such as physical deformity, growth restriction and delayed puberty. Children's physical health is also impacted by complications such as cardiac failure and arrhythmia, liver disease and endocrine disorders [10]. Thalassaemia also affects the children’s psychosocial health represented by emotional, social and school functioning [11-15].

As there are limited studies focused on HRQoL of thalassaemia among paediatrics patients, thus this current study brings forward to review the factors affecting the quality of life among thalassaemia children.

Methods

A comprehensive electronic search was conducted by using PubMed, Google Scholar and Science Direct. The search was limited to those articles written in English language and by using Pediatrics Quality of Life Inventory (PedsQL™) 4.0 generic core scale questionnaire only.

The 23-item PedsQL 4.0 encompasses the essential core domains for paediatrics HRQoL measurement: Physical Functioning, Emotional Functioning, Social Functioning and School Functioning [16]. The PedsQL requires the thalassaemia patients to recall the frequency of their problems occurred in the past month. Individual response options of which 5-point Likert scale from 0 (never a problem) to 4 (almost always a problem) was used for all items [16]. All items were then reverse-scored and linearly transformed into 0 to 100 point scale [16].

The Physical Health Summary Score is corresponding to the Physical Functioning scale score where it assesses the changes in
physical activities performed by individuals day to day. The Psychosocial Health Summary Score is the mean score result from the sum of items divided by the number of items completed in the Emotional, Social and School Functioning Subscales.

The emotional functioning measures the satisfaction, achievement of personal goals, personal controls, social interaction, self-concept and self-esteem [17] while the social functioning evaluates the existence of social relationship and activities [18, 19]. School functioning was used to assesses how many time a child absent from school due to illness and admitted to the hospital [20]. The Total Score is the sum of all the items over the number of items answered on all the scales. The Total Score indicates an overall measure of HRQoL, with higher points reflects the higher quality of life [21].

**Results**

After reviewing the titles and abstracts, a total of sixteen articles related to the factors affecting the quality of life of thalassaemia among paediatrics patients were identified. Of these, fourteen articles were on cross-sectional studies and the remaining articles were unmentioned designs (Table 1).

<table>
<thead>
<tr>
<th>Author(s)</th>
<th>Study Design and Sample Size</th>
<th>Factors Affecting Quality of Life</th>
<th>Factors Not Affecting Quality of Life</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sazlina et al (2015)</td>
<td>Cross-sectional study, 70 Malaysian thalassaemia patients</td>
<td>Age, Ethnicity, Educational level, Duration of thalassaemia, Types of thalassaemia, Presence of side effect from iron chelation treatment</td>
<td>Gender, Family income, Number of siblings with thalassaemia, Marital consanguinity, Frequency of blood transfusion and iron chelation treatment</td>
</tr>
<tr>
<td>M Ismail et al (2013)</td>
<td>Cross-sectional study, 75 Malaysian thalassaemia patients</td>
<td>Pre-transfusion haemoglobin level, Frequency of iron chelation treatment</td>
<td>Age, Ethnicity, Educational level, Types of thalassaemia, Types of chelation treatment</td>
</tr>
<tr>
<td>A Ismail et al (2006)</td>
<td>Cross-sectional study, 78 Malaysian thalassaemia patients</td>
<td>Age</td>
<td>Gender, Ethnicity, Family income</td>
</tr>
<tr>
<td>Thavorncharoensap et al (2010)</td>
<td>Cross-sectional study, 315 Thailand thalassaemia patients</td>
<td>Age at onset of anaemia before two years, Age at first transfusion before four years, Received blood transfusion, Severe condition</td>
<td>Gender, Frequency of blood transfusion, Types of thalassaemia, Complications, Serum ferritin level</td>
</tr>
<tr>
<td>Surapolchai et al (2010)</td>
<td>Cross-sectional study, 75 Thailand thalassaemia</td>
<td>Family income, Type of payment, Severe</td>
<td>Age, Gender, Educational level</td>
</tr>
<tr>
<td>Study</td>
<td>Design</td>
<td>Study Population</td>
<td>Variables Studied</td>
</tr>
<tr>
<td>------------------------------</td>
<td>----------------------</td>
<td>-----------------------------------</td>
<td>-----------------------------------------------------------------------------------</td>
</tr>
<tr>
<td>Torcharus et al (2011)</td>
<td>Cross-sectional study, 49 Thailand thalassaemia patients</td>
<td>Condition, Age at diagnosis, Received blood transfusion and iron chelation treatment</td>
<td>Age, Severe condition, Pre-transfusion haematocrit level</td>
</tr>
<tr>
<td>Sultana et al (2016)</td>
<td>Cross-sectional study, 266 Indian thalassaemia patients</td>
<td>Unmentioned result</td>
<td>Age, Gender, Frequency of blood transfusion</td>
</tr>
<tr>
<td>Sharma et al (2016)</td>
<td>Cross-sectional study, 75 Indian thalassaemia patients</td>
<td>Age, Gender, Socio-economic status</td>
<td>Unmentioned result</td>
</tr>
<tr>
<td>Dhirar et al (2016)</td>
<td>Cross-sectional study, 241 Indian thalassaemia children</td>
<td>Comorbid, Frequency of blood transfusion, Duration of treatment, Number of concomitant medications, Total number of hospital visits per year</td>
<td>Unmentioned result</td>
</tr>
<tr>
<td>Saha et al (2015)</td>
<td>Cross-sectional study, 365 Indian thalassaemia patients</td>
<td>Gender, Socio-economic status, Illiterate parents, Marital consanguinity, Family history of thalassaemia, Types of thalassaemia, Received blood transfusion, Pre-transfusion of Hb level</td>
<td>Unmentioned result</td>
</tr>
<tr>
<td>Jafari-Shakib et al (2017)</td>
<td>Cross-sectional study, 31 Iranian and Italian thalassaemia patients</td>
<td>Unmentioned result</td>
<td>Gender, Serum ferritin level, Baseline Hb level</td>
</tr>
<tr>
<td>Caocci et al (2012)</td>
<td>Cross-sectional study, 60 Middle Eastern thalassaemia patients</td>
<td>Delayed start of iron chelation therapy</td>
<td>Gender, Age at diagnosis, Age at first transfusion, Frequency of transfusion, Complications, Serum ferritin level</td>
</tr>
<tr>
<td>Tuysuz et al (2017)</td>
<td>Unmentioned design, 80 thalassaemia patients</td>
<td>Serum ferritin level, Complications</td>
<td>Unmentioned result</td>
</tr>
</tbody>
</table>
Several studies have reported age and gender as significant factors affecting the quality of life among children with thalassaemia [10, 15, 22-25]. Studies also found that ethnicity and educational level affect the quality of life among children with thalassaemia. It was reported in a cross-sectional study over one year conducted in Selangor, Malaysia [10]. However, another two studies in Malaysia did not document the same findings [11, 12]. Patients’ body mass index also does not give an effect towards the quality of life [26].

Worst total summary scores were found among thalassaemia children who had illiterate parents and their parents who married consanguinely [24]. Family income also had strong association with the quality of life among thalassaemia patients [24, 27]. However, both Malaysian studies revealed that family income seems not to have much effect towards the quality of life [11, 12].

Children who had family history of thalassemia both or either in maternal or paternal side had significantly better quality of life scores [24]. A study in Jordan indicated the family history of thalassaemia affects the quality of life [23]. Presence of comorbidity among thalassaemia children had poorer total quality of life scores as compared to those who did not [22, 28]. They also found that more number of visits to the hospital, the poorer the quality of life scores. Type of payment where the patients were covered by Universal Health Coverage Scheme was also associated with quality of life [27].

A study conducted among Middle Eastern children with beta thalassaemia found that age at diagnosis affects the quality of life [29]. However, inconsistent with these findings, a Thailand study reported that age at diagnosis did not give an effect towards the quality of life among thalassaemia children [27]. Type of thalassaemia had significant result towards the quality of life [24, 30]. However, two studies did not report the same findings [12, 15]. Duration of thalassaemia was found to be significantly related to HRQoL impairment [10, 22].

Regarding the clinical and treatment characteristics, age at onset of anaemia before two years [15, 22] and age at first transfusion before four years [15] were found to be significantly associated with HRQoL with low total summary scores. Types of treatment (blood transfusion and chelation treatment) had strong association with quality of thalassaemia patients’ life [10].

Also, thalassaemia patients who received the blood transfusion in previous past months were significantly low HRQoL scores [15, 24, 27]. For the frequency of blood transfusion and its duration of transfusion therapy, it had significant association with quality of life of thalassaemia patients [13, 24].
Thalassaemia patients who were received iron chelation therapy showed low quality of life score compared to who did not [22, 27]. The frequency of iron chelation treatment was found to be a factor that affects the quality of life, but its type of chelation treatment such as Desferrioxamine, Deferasirox, Deferiprone was not associated with quality of life [12]. Moreover, Caocci et al (2012) found that delayed start of iron chelation therapy and presence of side effect from chelation treatment such as pain at injection site, nausea and vomiting had an impact on quality of life among children with thalassaemia [10].

Besides, a Malaysian study documented that pre-transfusion of haemoglobin (Hb) level was a significant predictor of HRQoL scores [12]. The lower baseline of Hb affect the quality of life [27]. This study exposed that patients with Hb level higher than 9g/dL had significantly higher total summary scores than those patients with Hb level less than 9g/dL. Serum ferritin level did not show any association with quality of life among thalassaemia patients [15, 29, 31]. However, this was inconsistent with Boonchooduang and colleagues study.

Patients with severe condition (patients who age at onset of anaemia before two years old and age at first transfusion before four years old, pre-transfusion of Hb level less than 7mg/dL and diagnosed with homozygous beta thalassaemia) was significantly associated with low HRQoL in both Thailand studies [15, 27]. Assessment of the components of HRQoL showed patients having disease complication was the factor influencing the quality of life [23] but not in study by Thavorncharoensap et al (2010) and Caocci et al (2012).

Physical Health Summary Score

Physical Functioning
The studies in this review demonstrated links between quality of life and the physical functioning among thalassaemia children. Age of thalassaemia children, gender and socio-economic status were associated with physical functioning [25]. Children who had family history of thalassaemia either on maternal or paternal side had strong association towards physical functioning [23]. Besides, patients who covered by Universal Health Coverage Scheme was significantly associated with physical functioning [27].

Types of thalassaemia was associated with physical functioning [32]. In addition, age at onset of anaemia before two years and age at first transfusion before four years were significantly related to the physical functioning in a study conducted in Thailand [15] and India [22]. The physical functioning was also significantly associated with types of treatment [10] where thalassaemia patients who received regular blood transfusion during previous three months [15] and received iron chelation treatment [27] showed significant results. However, this finding was inconsistent with a study reported by M Ismail and colleagues (2013) where they found patients who were not on blood transfusion or iron chelation treatment were a predictor of poor physical health-related quality of life. Surapolchai et al (2010) and Dhirar et al (2016) documented the frequency of blood transfusion gave an impact towards physical functioning.

Furthermore, the presence of side effects from chelation treatment also identified as factors that affecting lower physical functioning of thalassaemia patients [10]. Pre-transfusion of Hb level and patients who
had low baseline Hb had significantly low HRQoL in physical dimension [12, 27]. Based on clinical characteristics, patients with early age at diagnosis before two years [27] and had disease complications [23] indicated significant results. Dhirar et al (2016) found treatment duration, number of concomitant medicines and presence of comorbidities were significantly associated with self-reported physical summary scores.

Out of all factors that were affecting the physical summary score, it was found that physical health summary score was significantly impacted by the mean annual serum ferritin level where the higher the serum ferritin level, the lower the physical quality of life score [30].

**Psychosocial Health Summary Score**

Regarding the socio-demographic details of thalassaemia patients, age was found to be related to psychosocial health summary score [15, 22]. A cross-sectional study found that patients with low family income or covered by Universal Health Coverage Scheme had significantly affected the quality of life than those children who did not [27]. While the psychological summary score was significantly result by the type of thalassaemia; the psychosocial quality of life in thalassemia intermediate was better than thalassemia major patients, and it was associated with diabetes where the presence of diabetes will worsen the HRQoL [30].

Duration of thalassaemia less than ten years impact the quality of life [10] same goes to pre-transfusion of Hb level where it significantly related to psychosocial health summary scores [12, 15]. Based on clinical characteristics, Surapolchai and colleagues (2010) study indicated that patients with severe condition, low baseline Hb, early age at diagnosis before two years and receive regular transfusion had significantly low HRQoL. The frequency of blood transfusion and iron chelation treatment also found to be significant factors that impact psychosocial health summary score [12]. Furthermore, psychosocial domain scores strongly associated with duration of treatment and number of visits to the hospital per year [22].

**Emotional Functioning**

Not all studies reported the significant factors affecting the quality of life of each emotional, social and school functioning among thalassaemia patients. The majority of the authors keep combined all of this functioning in one dimension which was psychosocial health summary scores since this three functioning act as domains in psychosocial health dimension. However, among all of the articles in this review, only one study reported the significant factors that affect emotional functioning among thalassaemia patients where they found emotional functioning was significantly associated with age, education level, duration of thalassaemia and types of treatment [10].

**Social Functioning**

There were significant association between social functioning with age, ethnicity, education level, duration of thalassaemia and types of treatment [10]. As in the previous study, Thavorncharoensap et al (2010) also found that age as a significant factor that affects lower social functioning. They also found pre-transfusion of Hb level, age at onset of anaemia before two years and age at first transfusion before four years, having a severe condition and received iron chelation treatment were significantly impaired HRQoL in school functioning subscale [15].
**School Functioning**

Based on a study among Malaysian children, school functioning was only associated with frequency of blood transfusion [10]. Age, pre-transfusion of Hb level, age at onset of anaemia before two years and age at first transfusion before four years and having a severe condition were significantly affect HRQoL [15]. Additionally, disease complications also associated with low quality of life [23].

**Discussion**

Most of the studies in this current review found that the total summary scale, physical health summary scores and psychosocial health summary scores were almost similar [10-12, 15, 23, 27, 29, 31-33]. The average of total scores in Malaysian studies approximately 69.0 [10, 11, 26]. These scores were lower when compared to both Thailand studies where the average scores were between 75.0 and 79.0 [15, 27].

The difference in the scores of HRQoL domains can be seen because most of the participants in Malaysian studies were homozygous beta-thalassaemia and transfusion dependent as compared to those participants in Thailand studies. Moreover, possible reasons were revealed that there were differences in cultures, experiences and perspectives between Thailand and Malaysia. Another study reported that the mean HRQoL score in adolescents was greater than 70, which indicated good HRQoL whereby they perceived themselves as being physically active and capable to perform normal regular activities [34]. Assessment on the subscales of psychosocial health summary score found that in most of the studies, the domain of school functioning scored the lowest except in a study by Jafari-Shakib et al (2017). Thalassaemia children need to undergo blood transfusions once every three to four weeks at day care centre or hospitals. Frequent school absenteeism for hospital visits, coping up with the school work and lack of energy when carry out academic activities may be additional problems, which affect the quality of life. However, this result was contrasted with other results in Egypt study where they found school functioning scores were better as the patients who need to transfuse their blood attended the clinic on Saturdays that were the schools’ day off [35].

The emotional dimension also affected because thalassaemia patients felt different from their peers. They attend to express negative beliefs about their lives and sometimes, they may felt sad, angry and hurt toward their illness. Children might have psychological and emotional troubles as early as the toddler stage. Some of them seek the ability to fulfil their needs on their own, however, their caregivers did not let them perform these tasks because of their illness. Thus, thalassaemia patients might develop shame and doubt about their ability to handle problems. Thalassaemia children, as they grew older, they were becoming more aware of themselves. Some of them become more responsible towards themselves and their illness. They were becoming more rational to share and cooperate, but thalassaemia prevented them from being more productive, and a sense of inferiority might develop instead and lower their emotional domain scores [15].

Furthermore, the treatment was emotionally demanding, where the blood transfusion and chelation therapy required repeated invasive procedures and hospital visits [36].

The low HRQoL in the physical domain in Jordan study documented that their thalassaemia children reported the presence...
of pain, aches and low energy when performing day to day activities [23]. Living with thalassaemia illness since early childhood causes many difficulties result in activity intolerance such as fatigue, general weakness and difficulty in breathing, which resulted from low Hb levels. Another reason probably because many patients did not come regularly for blood transfusion and thus, they would suffer from anaemic consequences that poorly impacted their physical activities and limited their exercise capacity.

Thalassemia children reported that they have no problems in socialising since the majority of the studies in this current review showed higher score compared to other domains. They had no troubles in getting along with other kids or being a member of a play team. This may be due to the non-appearance of evident disease complications at a young age, resulting in decreased feeling of stigmatisation [35].

**Total Summary Scores**

This review found that age was a significant predictor of HRQoL among thalassemia patients. Adolescent patients had significantly higher HRQoL than their younger counterparts where they experienced fewer symptoms of depression, reflecting a process of adjustment and coping with their illness [13].

Furthermore, complications and type of thalassaemia were not related to HRQoL reported in Thailand study. A small number of patients having complications in their study could explain this situation. Although patients who diagnosed with homozygous beta-thalassemia were predictable to have significantly low HRQoL since they were transfusion-dependent, but non-significant relationship was found. Similarly, a small number of homozygous beta thalassemia patients possibly explained this condition.

Presence of comorbidity among thalassaemia children had poorer total quality of life scores as compared to those who did not. Comorbidities affect severely both the physical and psychological quality of life where its absence enhances the overall quality of life. The number of comorbidities was a strong predictor of the poor physical and total quality of life score [22]. Frequent visits to the hospital have a negative impact on children's lives regarding the physical burden, psychological burden and school attendance, thus affecting their quality of life. Children have a constant stress of travelling all the way from long distances and being subjected to painful investigations and transfusion procedures.

Consistent with the findings from several studies, thalassaemia patients who received the blood transfusion and patients who received frequent blood transfusion having lower health score when compared with patients receiving less frequent. The patients typically have to undergo for blood transfusions once a month depends on the severity of the illness. The attendance for an entire day at the hospital leads to school absenteeism and might impaired HRQoL indirectly. In contrast, there were a few studies reported no significant relationship between the frequency of blood transfusion and HRQoL. This non-significant finding could be because of the questions used to assess HRQoL of the patients were related to the feelings and conditions of the patients during the previous month; therefore the number of transfusions per year might not be relevant to the HRQoL score.

This review also showed that frequency of iron chelation treatment was significantly related to HRQoL impairment. A frequent
Chelation therapy may lead to an increment in iron losses which indirectly influence their quality of life. In addition, delayed start of iron chelation also had an impact on total PedsQL. Delayed iron chelation can lead to excessive accumulation of iron in body organs. Health issues arise especially when excess iron is stored in the heart, liver and pancreas [29].

Additionally, pre-transfusion Hb level was found to be significant in HRQoL. The study revealed that patients with Hb level higher than 9 g/dL had a significantly higher HRQoL than those with Hb level less than 9 g/dL. This could explain by the fact that low Hb level linked with several symptoms, such as fatigue, general weakness and decreased mental alertness, which might impaired HRQoL of the patients [37]. Pre-transfusion Hb level should be supervised routinely to retain an optimal level of 9 to 10.5 g/dL [38]. In the case of blood transfusion, one possible explanation for the significant relationship between receiving a blood transfusion and low HRQoL is that patients who received blood transfusions during the three months prior to HRQoL assessment were those with low pre-transfusion Hb levels.

Baseline Hb levels did not show any association with quality of life of thalassaemia patients. Appropriate control of the disease in the patients may be the reason for almost acceptable quality of life where more than half of thalassaemia patients had Hb level greater than 8g/dL. Small sample size also could be the reason for this finding. Serum ferritin level also does not show any association with the quality of life of the patients. This inconsistent result possibly because of damage, the long-term iron overload occurs gradually; so short-term iron overload, as represented by an elevated serum ferritin level, did not cause significant visible symptoms or complications, and hence had no impact on their HRQoL.

**Physical Health Summary Scores**

**Physical Functioning**

The physical health summary score is based on the physical functioning, which assessed the level of physical activity and energy level of the participants’ over previous months. Most of the patients in the studies were 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. Thus, this might affect negatively their physical functioning.

Combined oral and subcutaneous iron-chelation therapy was negatively predicted the physical health, due to the burden of nightly subcutaneous injections of desferrioxamine and daily oral deferiprone tablet. Severe cases where the patients with transfusion dependence and iron overload probably need combined iron-chelation therapy to achieve serum ferritin levels below 1,000 to 1,500 ng/mL [39], a threshold that is known to be the most reachable tool and related with a reduced risk of iron overload-related complications such as heart failure in patients with thalassemia.

Presence of side effects from chelation treatment also affects physical health of thalassaemia patients. The burden of subcutaneous injections of iron chelation treatment five to seven days a week was associated with impaired HRQoL [15]. Furthermore, it is known that among the common side effect of chelation treatment are the pain at the injection site, nausea and vomiting. This side effect may contribute to
the reduced scores in this study as most of the patients received five or more times in a week of chelation treatment [27].

Since the severity of thalassaemia and treatment were not always associated with reductions in the HRQoL, a Thailand study by Surapolchai et al (2010) is the first report presenting a significant relationship between the HRQoL and family financial impact in thalassaemia patients. Surprisingly, type of payment had an impact on both children and parents’ perspectives. Self-payment could be implied to be higher household income and affordable medical expense; which assisted to predict the better parent-rated HRQoL.

Psychosocial Health Summary Scores

Predictor of lower psychosocial health summary score was the duration of thalassaemia less than ten years [10]. In the present study, those with duration of thalassaemia were those patients aged less than 13 years old. The younger patients would have fewer experiences in dealing with their illness and may have trouble to understand the disease they have, which could affect them emotionally. Moreover, in this study, most of the Malaysian children aged less than 13 years were on blood transfusion. Consequently, their social and school functions also affected as they need to be away from school to receive the transfusion or any other treatment. Hence, this could affect their psychosocial health. The older children could have more knowledge about thalassaemia, and they were able to understand the support from their parents, carers or peers to allow them to cope better.

Regarding the emotional functioning, adolescent males were found to have better emotional dimension than adolescent females [34]. Peer influence can be the reason for poor emotional functioning in female adolescents. Bullying and teasing due to the physical appearance probably affect the individuals’ self-esteem resulting in poorer HRQoL [40]. Sociocultural pressures such as family influences may also directly have an impact on the HRQoL of the adolescent's girls resulting in poor overall HRQoL [40].

When looking at subdomains of psychosocial health dimension, patients who received the blood transfusion and iron chelation treatment affect school functioning. Thalassaemia children need to require the blood transfusion to meet body demands. Literature reviews discovered that patients who received the blood transfusion related with school functioning dimension. It may be due to multiple visits to hospitals where these patients need to transfuse their blood. For several patients, maybe they had same complications after they get the blood transfusion. So in that case, they need to admit into the ward for more days.

Conclusion

Age, severe types of thalassemia, received of blood transfusion, serum ferritin level, frequency and side effects of iron chelating treatment, and presence of complications are the most documented contributory factors for low quality of life in thalassemia patients. This review gives an emerging knowledge regarding contributory factors that has a big impact in clinical practice in order to improve thalassemia patients’ quality of life. This serves as a foundation for the development of an effective preventive strategy via screening of thalassemia carrier among pre-marital adults as well as school students.

Acknowledgement
This study was funded by Short Term Grant by USM: 304/PPSP/6315004.

References


[18] Bowling A. Health-related quality of life: A discussion of the concept, its use and measurement background: The ‘Quality of Life’ Presented to the Adapting to Change Core Course, September 1999.


