Assessment of quality of life in transfusion dependent thalassemic children - need to address parents/care givers

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ABSTRACT

Background: Thalassemia is an autosomal recessive congenital disease caused by the reduced or absent beta globin chain synthesis of hemoglobin tetramer. The degree of imbalance between alpha and non alpha globin chains determines the severity of clinical manifestations. The disabling nature of the disease and chronic therapy affects the normal life causing psychosocial burden. Overall patient’s life, such as education, free-time, physical activities, skills, capabilities, and family adjustment is affected. The effects of which often result in psychological, emotional and social compromise. Health-Related Quality of Life (HRQoL) measurement is a multidimensional concept that focuses on the impact of the disease and its treatment on the well being of an individual.

Methods: A descriptive observational hospital based study was conducted over a period of 3 months. Transfusion dependency in thalassemic children aged between 5 years and 18 years was the inclusion criteria. Thalassemic children having debilitating illnesses unrelated to thalassemia were excluded. Quality of life was assessed using Pediatric Quality of Life Inventory (PedsQL™ 4.0)4. The tool assesses the quality of life in five domains: physical functioning (PF: 8 items), psychosocial functioning (sum of emotional, social and school functioning), emotional functioning (EF: 5 items), social functioning (SF: 5 items) and school functioning (SC: 5 items).

Results: Total of 125 children were enrolled with a mean age of 9.4±4.6 years (age range 5-18 yrs). According to the PedsQL questionnaire, the quality of life was similarly assessed by both parents and children. The total mean QoL score of the parents was 72.36±11.47 and of the children was 77.63±14.17. Emotional, school and psycho-social function were significantly affected according to both child and parents without statistical significance.

Conclusions: Thalassaemia patients and their parents require lifelong psychological support for prevention of mental health issues. By increasing the awareness and knowledge levels of the parents, we can help sick children in developing countries to get the best care locally and to thus improve HRQoL.

Keywords: HRQoL, Mental health issues, Transfusion dependent thalassemia

INTRODUCTION

Thalassemia is an autosomal recessive congenital disease caused by the reduced or absent beta globin chain synthesis of hemoglobin tetramer. The degree of imbalance between alpha and non alpha globin chains determines the severity of clinical manifestations.1 Thalassemia has its origin from mediterranean countries and is now present worldwide with high prevalence in mediterranean, middle east and central Asian countries.2 According to various reports, 7500-12,000 β-thalassaemia major babies would be born in India each year.3,5 Children with transfusion dependent beta thalassemia present with progressive pallor, failure to
thrive and splenomegaly. Regular blood transfusions and
iron chelation to counter iron overload state secondary to
blood transfusions form the mainstay of treatment. Bone
marrow transplant remains the curative treatment. Hospital
appointments or admissions for regular monthly
blood transfusion and/or treatment of complications make
children to skip school leading to school absenteeism
compromising their self-identity and making them
increasingly dependent on others. The disabling nature
of the disease and chronic therapy affects the normal life
causing psychosocial burden. Overall patient’s life, such
as education, free-time, physical activities, skills, capabilities, and family adjustment is affected. The
effects of which often result in psychological, emotional
and social compromise. Health-Related Quality of Life
(HRQoL) measurement is a multidimensional concept
that focuses on the impact of the disease and its treatment
on the well-being of an individual. The measures are seen
as ways of capturing patients’ perspectives of their
disease and treatment, their perceived need for health care
and their preferences for treatment and disease
outcomes. Various studies have shown that all patients
with thalassemia should undergo QOL assessment so that
interventions focused on the affected domain can be
implemented. Given the paucity of research in this area
and the need to rely on additional evidence-based data to
further improve patient care, we did a pilot study to
evaluate the quality of life in children with transfusion
dependent thalassemia attending our centre. Considering
that previous studies of childhood illness have shown that
parents ratings of their child’s HRQoL tend to be lower
and possibly reflect parental distress. We also
evaluated the differences between HRQoL reports of the
children and their parent-proxy.

METHODS
A cross sectional observational hospital based pilot study
Study was conducted for the period of 3 months from
June 2019 to September 2019 were included in the study.

Inclusion criteria
Transfusion dependency in consented thalassemic
children aged between 5 years and 18 years.

Exclusion criteria
Thalassemic children having debilitating illnesses
unrelated to thalassemia.

Quality of life was assessed using Pediatric Quality of
Life Inventory (PedsQL™ 4.0). The tool assesses the quality of life in five domains: physical (PF: 8 items), psychosocial (sum of emotional, social and school functioning), emotional (EF: 5 items), social (SF: 5 items) and school (SC: 5 items).

Each item is rated on a 5-point Likert scale from 0
(never) to 4 (almost always) and items are reversed
scored and linearly transformed to a 0-100 scale as
follows: 0=100, 1=75, 2=50, 3=25, 4=0.

Data were computed and analyzed by and SPSS
(Statistical Package for the Social Sciences) program
version 19.0. General characteristics of the patients
were presented in terms of percentage, mean, and standard
deviation and median for data not normally distributed.
For QoL, both total HRQoL score and physical,
emotional, social, school achievement and psychological
scores were presented in terms of mean and standard
deviation.

RESULTS
One hundred twenty five transfusion dependent
thalassemia children were included between the age
group of 5-18 years. Male babies outnumbered female
babies (male:female :: 73:52). Children were divided into
three age groups 5-7 yrs, 8-12 yrs, 13-18 yrs. 34 babies
contributed to 5-7 yrs age group, we had 51 babies in
8-12 yrs, 40 babies in 13-18 yrs.

Table 1: Demographic characteristics of the
participants (n=125).

<table>
<thead>
<tr>
<th>Variable</th>
<th>Value</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Gender</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>73</td>
<td>58.4</td>
</tr>
<tr>
<td>Female</td>
<td>52</td>
<td>41.6</td>
</tr>
<tr>
<td><strong>Age group (years)</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Young children</td>
<td>34</td>
<td>27.2</td>
</tr>
<tr>
<td>5-7</td>
<td></td>
<td></td>
</tr>
<tr>
<td>8-12</td>
<td>51</td>
<td>40.8</td>
</tr>
<tr>
<td>13-18</td>
<td>40</td>
<td>32.0</td>
</tr>
<tr>
<td><strong>Residence</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Urban</td>
<td>75</td>
<td>60.0</td>
</tr>
<tr>
<td>Rural</td>
<td>50</td>
<td>40.0</td>
</tr>
</tbody>
</table>

On recording the quality of life score by children and
parents, following results were obtained. Mean physical
score according to child was 81.57±16.33, parent
recorded 77.63±16.44 (p=0.45). For the social score
was 73.68±13.1 in children, parents recorded 68.42±14.04
(p=0.23). Emotional score as per children was
70.63±16.44, parents reported score was 66.68±15.1
(p=0.40). Score in school domain in children was
67.1±16.77, parents reported score was 60.52±15.17
(p=0.19). Regarding psycho social domain, Child
reported score was 65.78±12.38, parent reported score
was 63.15±12.82 (p=0.52). Total score was 77.63±14.17
in children and parents recorded score was 72.36±11.47.
Physical and social domains were significantly higher
reported by both child and parents as well as total
HRQoL score, though statistically insignificant.
Emotional, school and psycho-social function were
significantly affected according to both child and parents
without statistical significance.
Table 2: Children and parents total PedsQL score and single scale scores comparison.

<table>
<thead>
<tr>
<th>Questionnaire</th>
<th>PedSQL Questionnaire version</th>
<th>Mean</th>
<th>SD</th>
<th>Wilcoxon test</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total /Sum</td>
<td>Child</td>
<td>77.6316</td>
<td>14.17827</td>
<td>Z= -1.25</td>
<td>0.21</td>
</tr>
<tr>
<td></td>
<td>Parent</td>
<td>72.3684</td>
<td>11.47079</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Physical</td>
<td>Child</td>
<td>73.6842</td>
<td>13.10663</td>
<td>Z=-1.19</td>
<td>0.23</td>
</tr>
<tr>
<td></td>
<td>Parent</td>
<td>68.4211</td>
<td>14.04879</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Emotional</td>
<td>Child</td>
<td>77.6316</td>
<td>16.44591</td>
<td>Z=-0.83</td>
<td>0.40</td>
</tr>
<tr>
<td></td>
<td>Parent</td>
<td>73.6842</td>
<td>13.10663</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Social</td>
<td>Child</td>
<td>81.5789</td>
<td>16.33441</td>
<td>Z= -0.75</td>
<td>0.45</td>
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<tr>
<td></td>
<td>Parent</td>
<td>77.6316</td>
<td>16.44591</td>
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<td></td>
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<tr>
<td>School</td>
<td>Child</td>
<td>67.1053</td>
<td>16.77596</td>
<td>Z= -1.29</td>
<td>0.19</td>
</tr>
<tr>
<td></td>
<td>Parent</td>
<td>60.5263</td>
<td>15.17442</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Psychosocial function</td>
<td>Child</td>
<td>65.7895</td>
<td>12.38987</td>
<td>Z= -0.65</td>
<td>0.52</td>
</tr>
<tr>
<td></td>
<td>Parent</td>
<td>63.1579</td>
<td>12.82473</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

DISCUSSION

In the present study, assessing the QoL in transfusion dependent thalassemia children showed the impact of transfusion dependent thalassemia on the patients performance in different aspects of life.

Psychosocial performance was the significantly affected domain. Followed by school and emotional domains. Similar reports were found in study by Sachith Mettananda, Adriana Ismail.12,13

On comparing parents and child self reporting, it was observed that parents score was low compared to child’s score.

It would appear that parents tend to consider HRQoL of their children more compromised in domains dealing with interpersonal relationships (Social) rather than those concerning physical impairment. i.e., Physical functioning is better than social function. Similar results were seen in study by Surapolchai P, Caocci et al.14,2

A possible explanation may be that parents of thalassemic children unconsciously project their pessimistic feelings onto their children’s functioning.

Low school domain score signifies the role of healthcare providers, counsellors and school teachers in helping children to overcome this problem.

CONCLUSION

Our pilot study showed the difference in self reporting of child and parent proxy reporting. It highlighted the significant effect of transfusion dependent thalassemia on psychosocial, school and emotional domains. Parent ratings of their child’s HRQoL were lower for emotional functioning, psychosocial functioning and school functioning, suggesting the need to enhance the understanding and support of the parents. By increasing the awareness and knowledge levels of the parents, we can help sick children in developing countries to get the best care locally and to thus improve HRQoL.

Limitations of our study were small sample size, we obtained HRQoL measurements in a cross-sectional manner we did not perform a complete cognitive or psychological evaluation based on normative scale scores, which would have been helpful in order to avoid confounding factors (i.e. cognitive, psychological or psychiatric problems in parents or children).

We conclude that Thalassaemia patients and their parents require lifelong psychological support for prevention of mental health issues. Several effective psychological strategies are available. Cognitive-Behavioural Family Therapy (CBFT) can be an effective psychological approach to children with beta-thalassaemia major, capable of increasing compliance to treatment, lessening the emotional burden of disease and improving the quality of life of caregivers as reported by Mazzone L et al.

Overall, HRQoL measurement should be part of therapy in children with chronic diseases and appropriate intervention can improve the quality of life of children and care givers.

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REFERENCES


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