An epidemiologic assessment on the oral manifestations of B thalasemia in-patients referred to oncology wards in Tehran during 2000-2001

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ABSTRACT

Purpose: B thalasemia had been reported as a highly, prevalent genetic disorder in the Middle East region associated with some dental and periodontal complications. This study was designed to evaluate the frequency of dental and periodontal problems in thalasemic sufferers.

Materials & Methods: A group of 100 B thalasemic patients referred to the Hematology wards in selected university hospitals in north eastern region of Tehran were investigated for rise in upper lip, promineny of the cheek, pale mucosa, premaxillary expansion, crudely saddled nose, dental caries rate and periodontal status. Data were recorded and rates were compared with their percentile.

Results: Early diagnose of the disease was shown to play important introductions of future complications including orofacial region. This was due to on time commencement of the blood transfusion leading to a more natural growth.

Conclusion: Appropriate and on time blood transfusions in thalasemic children could clearly prevent many of the future orofacial complications.

Keywords: Oral manifestations, B thalastemia, Oncology.

INTRODUCTION

There are several parts in maxillofacial complex, which would be affected in thalasemic patients, including maxilla and upper lip, teeth position, periodontium, nose, cheek and skull. Early diagnosis of the disease is considered as an advantage towards prevention of such changes in the facial region. Dentist’s knowledge could lead him to a more appropriate diagnosis associated with an on time treatment. Lack of efficient hemoglobin is the main finding in thalasemic cases which is associated with insufficient and imbalanced accumulation of the globulin chains. Imbalances in globulin synthesis are seen in B thalasemia leading to hemolysis with varying degrees. Patients suffering from thalasemia major look severely pale after a few months. Enlargement of the liver and spleen are also seen associated with a clear delay in new born weighting and development. These cases will usually get worse by time instead of the cure.

Without blood transfusion, hemoglobin level could drop to 3-5 gr./dl. Most of the thalasemic cases are seen with smaller sized body, a larger head, with a coolies face. Maxillary and mandibular over growth is clearly seen particularity in the vertical dimension. This investigation was designed to evaluate the facial changes as well
as crowding, and mucosal changes in a group of thalasemic patients residing in Tehran.

MATERIALS & METHODS

This descriptive study performed through observation and interview method on a group of 100 individuals (44 males and 56 females). Patients aged 4-25 years were selected from those referred to the hematology wards in five major general hospitals in Tehran for blood transfusion. A thorough clinical examination including both intra and extra oral assessments were carried out as routine. Collected data were recorded from each observation of the shape of the face, occlusion, gingival status and the soft tissue color, in the forms provided for this purpose. Data analysis was performed using a Chi-square test.

RESULTS

Mean age of the patients was 13.82 years with the majority lying between 10-15 years. Most of the cases were at their late primary or early high school educational stage. Parents were mainly reported to have little or no education (24%) and only 8% of the mothers had finished their high school.

It was of note that 93% of the patients had a family history of the disease with a sister being involved in most instances. Hemoglobin rate was at 7.48gr/dl (+0.72) in the whole population of the study (Table 1).

The most frequently diagnosed age for the disease was found to be 2-4 years of age with the mean diagnosis age being at 2.12 (+0.93) years (Fig1).

Most of the patients had reported to have received the first dose of Decaferal injection at the age of 5 (15%) (Fig2).

Table 1. Distribution of thalasemic patients with varying levels of hemoglobin.

<table>
<thead>
<tr>
<th>Hemoglobin rate</th>
<th>No</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>&gt;6/9</td>
<td>14</td>
<td>14</td>
</tr>
<tr>
<td>7-7/9</td>
<td>39</td>
<td>39</td>
</tr>
<tr>
<td>8-8/9</td>
<td>45</td>
<td>45</td>
</tr>
<tr>
<td>≥9</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Total</td>
<td>100</td>
<td>100</td>
</tr>
</tbody>
</table>

Table 2. Blood Transfusion starting age among the thalasemic cases of this study.

<table>
<thead>
<tr>
<th>Starting age for blood transfusion</th>
<th>No</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>&gt;1 Year old</td>
<td>13</td>
<td>13</td>
</tr>
<tr>
<td>1</td>
<td>12</td>
<td>12</td>
</tr>
<tr>
<td>2</td>
<td>18</td>
<td>18</td>
</tr>
<tr>
<td>3</td>
<td>11</td>
<td>11</td>
</tr>
<tr>
<td>4</td>
<td>14</td>
<td>14</td>
</tr>
<tr>
<td>5</td>
<td>12</td>
<td>12</td>
</tr>
<tr>
<td>6</td>
<td>5</td>
<td>5</td>
</tr>
<tr>
<td>7</td>
<td>7</td>
<td>7</td>
</tr>
<tr>
<td>8t</td>
<td>8</td>
<td>8</td>
</tr>
<tr>
<td>Total</td>
<td>100</td>
<td>100</td>
</tr>
</tbody>
</table>
It was also noted that the dental caries was a problem in most of the patients (97%) with only 12% of them also suffering from a type of diabetes too.

Other systemic diseases related to the cases are listed in table 3.

**Table 3. Distribution of thalasemic patient’s base on their systemic disease.**

<table>
<thead>
<tr>
<th>Systemic disease</th>
<th>No</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cardiac disease</td>
<td>20</td>
<td>20</td>
</tr>
<tr>
<td>Headache, Nausea</td>
<td>80</td>
<td>80</td>
</tr>
<tr>
<td>Liver disease</td>
<td>15</td>
<td>15</td>
</tr>
<tr>
<td>Diabetes</td>
<td>12</td>
<td>12</td>
</tr>
<tr>
<td>Developmental disturbances</td>
<td>58</td>
<td>58</td>
</tr>
<tr>
<td>Fatigue weakness</td>
<td>67</td>
<td>67</td>
</tr>
<tr>
<td>Bone fracture</td>
<td>34</td>
<td>34</td>
</tr>
<tr>
<td>Joint pain</td>
<td>74</td>
<td>74</td>
</tr>
<tr>
<td>Backache</td>
<td>60</td>
<td>60</td>
</tr>
<tr>
<td>Dental caries</td>
<td>97</td>
<td>97</td>
</tr>
<tr>
<td>Psychologic complications (Depression)</td>
<td>60</td>
<td>60</td>
</tr>
</tbody>
</table>

In total, 40% of the cases had spleenectomy due to their complications.

Overall 78% of the patients represented a Mongoloid face with 42% of them showing some degrees of crowding in both jaws (table 4).

Sixty percent of the cases were scored poor for their oral hygiene index with the remaining 40% being at an acceptable level (table 5).

Chi-square test showed a significant difference on the maxillary protrusion level between those who received on time blood transfusion and those who had it in a later stage (P<0.001, table 6).

Further findings indicated that those who had received the first blood transfusion at the age of 5 suffer from a higher degree of crowding compared to those who started it later, with a level of significance (P<0.005) (table 7). However, any logic relationship needs to be looked at.

It also mentioned that early blood transfusion was less happening in whom had started it at the end of the first year of life.

**Table 4. Distribution of maxillofacial anomalies among thalasemic sufferers.**

<table>
<thead>
<tr>
<th>Maxillofacial Disorders</th>
<th>Yes</th>
<th>No</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>number</td>
<td>Percent</td>
<td>number</td>
</tr>
<tr>
<td>Prominent Zygote (Zygomatic Exostosis)</td>
<td>69</td>
<td>69</td>
<td>31</td>
</tr>
<tr>
<td>Retruded Nose</td>
<td>66</td>
<td>66</td>
<td>34</td>
</tr>
<tr>
<td>Achrocephalic skull</td>
<td>57</td>
<td>57</td>
<td>43</td>
</tr>
<tr>
<td>Mongoloid Face</td>
<td>78</td>
<td>78</td>
<td>22</td>
</tr>
<tr>
<td>Lifted upper Lip</td>
<td>59</td>
<td>59</td>
<td>41</td>
</tr>
<tr>
<td>Maxillary enlargement</td>
<td>53</td>
<td>53</td>
<td>47</td>
</tr>
<tr>
<td>CI II malocclusion</td>
<td>50</td>
<td>50</td>
<td>50</td>
</tr>
<tr>
<td>Pale mucosa</td>
<td>70</td>
<td>70</td>
<td>30</td>
</tr>
<tr>
<td>Crowding</td>
<td>42</td>
<td>42</td>
<td>58</td>
</tr>
</tbody>
</table>
Table 5. Distribution of periodontal disease among thalasemic patients.

<table>
<thead>
<tr>
<th>Periodontal status</th>
<th>Good</th>
<th>Moderate</th>
<th>Poor</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Number</td>
<td>%</td>
<td>Number</td>
<td>%</td>
</tr>
<tr>
<td>Gingival status</td>
<td>34</td>
<td>34</td>
<td>31</td>
<td>31</td>
</tr>
<tr>
<td>Oral hygiene status</td>
<td>40</td>
<td>40</td>
<td>22</td>
<td>22</td>
</tr>
</tbody>
</table>

Table 6. Distribution of maxillary protrusion among thalasemic sufferers in relation to blood transfusions age.

<table>
<thead>
<tr>
<th>Lifted upper lip</th>
<th>Yes</th>
<th>No</th>
<th>Age</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Number</td>
<td>%</td>
<td>Number</td>
</tr>
<tr>
<td>&lt; 1 Year old</td>
<td>2</td>
<td>15.4</td>
<td>11</td>
</tr>
<tr>
<td>1</td>
<td>3</td>
<td>25.0</td>
<td>9</td>
</tr>
<tr>
<td>2</td>
<td>6</td>
<td>33.3</td>
<td>2</td>
</tr>
<tr>
<td>3</td>
<td>8</td>
<td>72.7</td>
<td>3</td>
</tr>
<tr>
<td>4</td>
<td>13</td>
<td>91.2</td>
<td>1</td>
</tr>
<tr>
<td>5</td>
<td>11</td>
<td>91.7</td>
<td>1</td>
</tr>
<tr>
<td>6</td>
<td>4</td>
<td>80</td>
<td>1</td>
</tr>
<tr>
<td>7</td>
<td>6</td>
<td>85.7</td>
<td>1</td>
</tr>
<tr>
<td>8+</td>
<td>6</td>
<td>75</td>
<td>2</td>
</tr>
<tr>
<td>Total</td>
<td>59</td>
<td>59</td>
<td>41</td>
</tr>
</tbody>
</table>

Table 7. Distribution of crowding in thalasemic sufferers in relation to blood transfusions age.

<table>
<thead>
<tr>
<th>Crowding</th>
<th>Yes</th>
<th>No</th>
<th>Age</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Number</td>
<td>%</td>
<td>Number</td>
</tr>
<tr>
<td>&lt; 1 Year old</td>
<td>2</td>
<td>15.4</td>
<td>11</td>
</tr>
<tr>
<td>1</td>
<td>3</td>
<td>16.7</td>
<td>10</td>
</tr>
<tr>
<td>2</td>
<td>7</td>
<td>38.9</td>
<td>11</td>
</tr>
<tr>
<td>3</td>
<td>3</td>
<td>27.3</td>
<td>8</td>
</tr>
<tr>
<td>4</td>
<td>16</td>
<td>42.9</td>
<td>8</td>
</tr>
<tr>
<td>5</td>
<td>10</td>
<td>83.3</td>
<td>2</td>
</tr>
<tr>
<td>6</td>
<td>3</td>
<td>60</td>
<td>2</td>
</tr>
<tr>
<td>7</td>
<td>4</td>
<td>57.1</td>
<td>3</td>
</tr>
<tr>
<td>8+</td>
<td>6</td>
<td>62.5</td>
<td>3</td>
</tr>
<tr>
<td>Total</td>
<td>42</td>
<td>42</td>
<td>58</td>
</tr>
</tbody>
</table>

Malocclusions was also seen to be more happening in cases with blood transfusion as late as 8 years of age. In the case of periodontal disease, patients who started to have blood transfusion after the age of 5 years showed a greater chance of the disease involvement (table 7).
DISCUSSION

There are several reports on the issue of thalasemia and its relation to dentofacial problems. (5) As most of the involved children received some sort of blood transfusion the effect of early or late transfusion on dentofacial anomalies had been investigated. (6)

It is believed that promoting health education could directly cause an obvious reduction in the number of thalasemic children. This has been more documented in children under the age of 10 years. (6) These children require regular dental attendance, which could help them with having simple early treatments instead of invasive works. In this regards, findings of the present study showed that inefficient and delayed blood transfusion is a case to notice in this group of children, which could automatically lead to dentoskeletal deformities. (5) Headache and dizziness were seen to be associated with dental caries in almost all patients. Desantis (1998) stated that 67% of the thalasemic patients observed were suffering from atypical diabetes. (7,8) It has also been suggested that the best time for starting the Disferal insertions being 3 years of age (3), disferal injections before this age could cause a reduction on growth speed and epiphysis distortion. (9) Results of the present investigation indicated that most of the patients who started Dispersal injection after 5 years of age, had represented side effects of thalasemia. These patients had also shown spleen problems similar to the findings of pervious reports of spleenectomy. (10) More than 1/3 of the population of this study had some degrees of periodontal disease reparations.

A deficient oral health care was seen in the group studied here similar to these controlled by Cappers (1994). (9) Vanarshi (2000) stated that thalasemic patients who were evaluated in their investigation were suffering from lung, gastrointestinal and skin infections. (5,10) Maxillofacial deformities were seen in 53% of the cases of this investigation a comparable to 90% maxillary enlargements reported earlier which could be explained with only enlargement being reported in those investigations. (3,4) Upper lips with high position was seen in almost similar number of cases compare to an earlier report (80% Compare to 90%). (5)

Half of the cases showed to have malocclusion with similarity to two earlier reports of 46% and 61%. (2,5)

CONCLUSIONS

As starting age for blood transfusion is critical in prevention of dental and periodontal diseases as well as some systemic complications this intervention is clearly recommended as early as possible.

REFERENCES


