ORAL AND DENTAL MANIFESTATIONS IN ADULTS WITH OSTEOGENESIS IMPERFECTA

Rønnaug SÆVES, DDS 1, Stefan AXELSSON, DDS, PhD 1, Lena L. WEKRE, MD 2, Kari STORHAUG, DDS, PhD 1

1 TAKO-Centre, Resource Centre for Oral Health in Rare Medical Conditions, Lovisenberg Diakonale Hospital Oslo, Norway
2 TRS - National Resource Centre for Rare Disorders, Sunnaas Rehabilitation Hospital, Norway

INTRODUCTION

Osteogenesis imperfecta (OI) is a genetic disease which involves connective tissues. In a Scandinavian material the prevalence is found to be 4-6 in 100,000 individuals. OI is characterised by bone fragility, joint laxity and occasionally dentinogenesis imperfecta (DI) as well as other dental abnormalities. About 50 % of individuals with OI exhibit DI type I according to the literature. The prevalence of DI is highest in OI types III and IV.

Class III malocclusion is reported in 67-80 % of individuals with OI. There is a high prevalence of agenesis of permanent teeth (10-18 %) in individuals with OI, (6-7 % in controls). Taurodontism is a morphologically abnormal tooth structure characterised by an apical extension of the pulp chamber. Taurodontism has been reported in permanent teeth (10-18 %) in individuals with OI, (6-7 % in controls).

STUDY GROUP AND METHODS

This study is part of an ongoing project describing a population of adults with OI in Norway, including individuals aged 25 to 100 years. The study group comprises 60 individuals (22 males, 38 females) with an age range from 27 to 83 years. We have estimated the number of individuals with OI within the investigated age group to be 150, and the present study group of 60 individuals is a representative selection.

Oral and dental abnormalities were assessed clinically, studied in orthopantomograms, lateral cephalograms, on dental study models and on intra- and extraoral photographs. Due to shortness of the neck it was not possible to obtain radiographs from all patients.

The dental examination included registration of DI, discoloration, missing and filled teeth, occlusion, attrition, and mandibular movement capacity. Data from the OI-group were compared with data from an epidemiological study in Norway (Schuller & Holst, 1998) and the mandibular movement capacity was compared with Helkimo (1974).

The participants were also interviewed about oral health, dental habits and temporomandibular joint problems.

The age and gender distribution is illustrated below.

RESULTS

Clinical

DI

In the study group 15 % had clinical and radiographic DI. DI has earlier been reported in 28-50 % of patients with OI and the prevalence of DI has been reported to be higher in the more severe types of OI.

Mandibular overjet was present in 13 % in the study group. This is significantly higher than in a general Scandinavian population (about 3 %).

DI

Range of mandibular movement

The mandibular movement capacity was not significantly different compared with the control group.

<table>
<thead>
<tr>
<th></th>
<th>Helkimo, 1974 (n=521)</th>
<th>Present study, 2005 (n=60)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Max. protrusion</td>
<td>2.3 ± 0.8</td>
<td>3.0 ± 1.9</td>
</tr>
<tr>
<td>Max. opening</td>
<td>44.7 ± 7.3</td>
<td>49.7 ± 7.0</td>
</tr>
<tr>
<td>Max. movement to right</td>
<td>9.3 ± 2.6</td>
<td>9.0 ± 2.1</td>
</tr>
<tr>
<td>Max. movement to left</td>
<td>9.3 ± 2.6</td>
<td>9.2 ± 2.1</td>
</tr>
<tr>
<td>Max. protrusion</td>
<td>8.2 ± 2.5</td>
<td>8.1 ± 1.9</td>
</tr>
</tbody>
</table>

Oral health

Missing teeth and filled teeth are two indicators used to describe general oral health. Individuals with OI in the middle-aged adult group (35-44 years) showed 2.2 missing teeth compared to 1.0 missing tooth in the control group. In the older adult age group (45-54 years) the study group showed 4.3 missing teeth compared to 2.6 in the control group. The number of filled teeth in the study group and in the control group were nearly equal in the middle-aged adult group (17.0 vs. 17.1) and in the older adult age group the differences were somewhat larger (15.9 vs. 18.0).

CONCLUSIONS

The study so far indicates low prevalence of clinical and radiographic DI compared to earlier studies. Mandibular overjet in 13 % is based on dentoalveolar abnormalities. A class III malocclusion, which most often is due to a skeletal discrepancy between jaws and the cranial base, is often masked by dental compensation and is more frequent than the variable “mandibular overjet”.

The oral health of adults with OI in Norway is good and the results indicate small differences with the exception of missing teeth compared with the Norwegian epidemiological study. A high percentage of individuals with OI use the dental services regularly, at least once yearly. This can be one factor affecting oral health positively.

REFERENCES