Dental features in patients with Turner syndrome

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ABSTRACT. Aim This was to investigate the dental characteristics (caries sensitivity and tooth crown size of permanent elements) in subjects affected by Turner Syndrome (TS). Patients and methods A group of 25 patients affected by TS, aged from 4 to 18 years, was selected and the data were compared to those of an age matched control healthy group. The caries index values in TS patients are higher in the permanent (6.4 vs. 3.9), mixed (0.5 vs. 0.75) and primary dentition (0 vs. 1). The mesio-distal diameter in TS patients was significantly reduced for every tooth measured (in particular for the lower first permanent molar). Results and conclusion This investigation confirms that numeric aberration of the X chromosome most likely affects the quantitative and qualitative excretion of amelogenin so that teeth often present enamel defects (reduced crown size and enamel hypoplasia). High caries index values (DMFT) highlight the demand of early preventive measures mostly focused on special care patients. Keywords: Turner syndrome, Crown width, Caries, Enamel hypoplasia.

Introduction

Turner syndrome (TS) is a symptom complex characterised by the presence of a single X functioning chromosome. TS occurs in 1:1500-2500 live female births.

The clinical signs depend on which genes cheat the deactivation. Anyway some characteristics are typical: short stature, gonadal dysgenesis and hormonal disorders (oestrogen deficiency), phenotypic features (congenital lymphedema, epicanthal fold, short and webbed neck, ptosis of eyelids, prominent ears).

The hormonal variation is responsible of the different cranio-facial growth pattern affecting the endocondral ossification of the skull structures.

TS patients are frequently carriers of congenital heart disease (aortic coarctation).

Mental development is usually regular even if psychological troubles have been observed.

Many studies about the aberration of X chromosome showed the relation between the sexual chromosome and dental growth. Anyway the effect of sexual chromosome aberration on dental morphology is less explored [Midtbø and Halse, 1994].

The most frequent oral findings in TS patients are: small teeth, short roots, narrow upper arch and high palatal vault, hypoplastic mandible.

The permanent dentition is characterised by a deviating pattern with respect to crown morphology; this anomaly could be responsible of increased frequency of occlusal anomalies [Midtbø and Halse, 1994; Alvesalo and Laine, 1992].

The aim of this study was to detect the dental features of a group of 25 patients with TS in order to formulate specific guidelines for dental prevention and therapy.

Patients and methods

Twentifive patients with TS (aged from 4 to 18 years) came from all over Italy for evaluation with the “Turner Syndrome Study Group” of the University Hospital of Bologna and were selected for the present study. They were compared to a control group composed by 25 healthy girls of the same age range randomly selected.

All the subjects underwent:
- dental examination;
- alginate impressions for study cast;
- x-ray examination (orthopantomography).

The number of teeth, enamel hypoplasia and the caries index (DMFT, dmft) were detected clinically according to the WHO method.
The mesio-distal permanent crown diameters were measured by means of a digital caliper on the dental casts according to Moorees [Grembowski et al., 1988]. Teeth not fully erupted or damaged by trauma, restorations and deep caries were excluded.

Dental anomalies regarding shape, number and position were detected on the orthopantomography.

The two groups were divided into three sub-groups according to the dentition:
- Turner/Control Sub-group A: 15 patients with permanent dentition;
- Turner/Control Sub-group B: 4 patients with mixed dentition;
- Turner/Control Sub-group C: 6 patients with primary dentition.

**Results**

The caries index was calculated separately for each sub-group and compared.

The Turner sub-group A showed a mean DMFT of 6.4 (D=3.8; M=3.5; F=1.7) while the Control sub-group A showed a mean DMFT 3.9 (D=2.5; M=1.7; F=3.9).

Mean caries index of the Turner sub-group B was 0.5 (D=0.3; M=0; F=0.3), while in the Control sub-group B it was 0.75 (D=0.3; M=0; F=0.5).

Mean dmft was 0 in the Turner sub-group C (D=0; M=0; F=0) and 1 in the Control sub-group C (D=0.7; M=0; F=0.3).

Enamel hypoplasia was detected in 10 Turner patients (40%): of 504 elements 31 were affected (6.1%). Among the control patients, only 3 of them showed enamel defects of 8 elements over 501 (1.5%).

In the study group the mesio-distal crown diameters of the permanent elements were reduced compared to the control group. The average diameter difference of the Turner patients is 1.4 mm: in particular the width of the crown of the first lower molar is 23.3% smaller.

The average difference between the teeth of the 2 groups is represented in table 1 and figure 1.

**Discussion**

The mesio-distal diameter is significantly reduced in the Turner Group: it is generally accepted that the dental crown morphology cannot be altered after full mineralisation has been reached. The critical period of odontogenesis is therefore between the initial development stage and the mineralisation stage of the dental crown [Midtbø and Halse, 1994]. The human enamel protein gene amelogenin is expressed from both the X and the Y chromosomes: it is supposed that chromosomal aneuploidy can directly influence enamel formation and crown size [Kollar, 1983; Lippe, 1990].

The reduced size of the tooth crown probably is function of the reduced thickness of the enamel layer. The effect of sex chromosome aberrations on dental morphology is less explored.

The tooth size is dependant on the number of X chromosomes: the more the Xs, the larger, mainly due to an increase in the enamel layer. It seems that genes on the X chromosomes control growth in general and that the X chromosome is able to influence stature, tooth size and enamel thickness.

**Table 1 - Mean tooth size of both groups.**

<table>
<thead>
<tr>
<th>Tooth</th>
<th>Turner</th>
<th>Control</th>
<th>Δ mm</th>
<th>Δ %</th>
</tr>
</thead>
<tbody>
<tr>
<td>+1+</td>
<td>7.3</td>
<td>8.3</td>
<td>1.0</td>
<td>12%</td>
</tr>
<tr>
<td>-1</td>
<td>4.7</td>
<td>5.6</td>
<td>0.9</td>
<td>16.1%</td>
</tr>
<tr>
<td>+2+</td>
<td>5.2</td>
<td>6.2</td>
<td>1.0</td>
<td>16.1%</td>
</tr>
<tr>
<td>-2</td>
<td>5</td>
<td>6</td>
<td>1.0</td>
<td>16.7%</td>
</tr>
<tr>
<td>+3+</td>
<td>6.9</td>
<td>8.1</td>
<td>1.2</td>
<td>14.8%</td>
</tr>
<tr>
<td>-3</td>
<td>5.7</td>
<td>7</td>
<td>1.3</td>
<td>18.6%</td>
</tr>
<tr>
<td>+4+</td>
<td>6.1</td>
<td>6.8</td>
<td>0.7</td>
<td>10.3%</td>
</tr>
<tr>
<td>-4</td>
<td>5.9</td>
<td>7</td>
<td>1.1</td>
<td>15.7%</td>
</tr>
<tr>
<td>+5+</td>
<td>5.5</td>
<td>6.7</td>
<td>1.2</td>
<td>17.9%</td>
</tr>
<tr>
<td>-5</td>
<td>6.2</td>
<td>7.2</td>
<td>1.0</td>
<td>13.9%</td>
</tr>
<tr>
<td>+6+</td>
<td>9.1</td>
<td>10.2</td>
<td>1.1</td>
<td>10.8%</td>
</tr>
<tr>
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<td>8.9</td>
<td>11.6</td>
<td>2.7</td>
<td>23.3%</td>
</tr>
<tr>
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<td>8.4</td>
<td>10.9</td>
<td>2.5</td>
<td>22.9%</td>
</tr>
<tr>
<td>-7</td>
<td>8.3</td>
<td>10.5</td>
<td>2.2</td>
<td>21.0%</td>
</tr>
</tbody>
</table>
X chromosome mapping allowed the location of the genes involved in the odontogenesis on the short arm (locus p 22): in particular AMGX is the gene encoding an enamel organic matrix protein excreted by the ameloblasts, which regulates the formation of crystallites during the secretory stage of enamel development [Salido et al., 1992; Sasaki and Shimokawa, 1995].

The prevalence of enamel hypoplasia in TS patients is higher than in Control group. Absence or aberration of one of the two X chromosomes can affect the enamel excretion, quantitatively (reduced tooth diameter) and qualitatively (enamel hypoplasia). Furthermore it is supposed that the reduced oxygen demand during the development of the dental buds, due to the congenital heart disease (30% of the Turner patients are affected by CHD), could influence the enamel structure so that enamel hypoplasia is more frequent.

Clinically the higher sensitivity to dental decay can be due to the enamel structural defects. As a matter of fact, DMFT is higher in TS patients than in the Control Group (6.4 vs. 3.9), D (decayed) and M (missing) values, higher than the F (filled), point out a difficult access to dental treatments.

The observed dmft=0 in patients with deciduous dentition is a remarkable finding, being pertinent to patients who had early access to our dental service. Early approach can be a very effective way to get parents and patients involved in the dental prevention system.

The operator, focusing on prevention rather than therapy, can explain the importance of oral hygiene and fluoride assumption (tablets, gel, varnish, milk, water, etc).

Conclusions

In conclusion, our investigation confirms the hypothesis that numeric aberration of the X chromosome most likely may affect the excretion of amelogenin quantitatively and qualitatively so that the dental elements often present enamel defects (reduced crown size and enamel hypoplasia).

High caries index values (DMFT) highlight the demand of early preventive measure mostly focused on special care patients.

Systemic fluoride assumption (to be started at 6 months of age) should be prescribed by paediatricians in order to minimise the caries risk along with oral hygiene practices and reduced sugar consumption.

References


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