

Dental Health Considerations & Solutions in Patients with Turner Syndrome

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Introduction

Females with Turner Syndrome (TS) encounter unique challenges in creating and maintaining oral and dental health. Because TS is associated with abnormalities in craniofacial structure and tooth development, a TS patient often functions with underdeveloped maxillofacial processes, misaligned bites, compromised teeth, a higher risk for tooth decay, and other challenges, while potentially limited fine motor skills can make caring for her teeth difficult. Because of these challenges, serving TS patients successfully requires diverse and creative solutions. This paper will address developmental issues and subsequent pathologies common to TS patients, and suggest potential solutions.

Dental health challenges related to Turner Syndrome

These oral conditions are typical of TS patients:¹

Developmental Pathologies

Abnormal oral development

- Asymmetry of facial bones
 - Retrognathia
 - Mandibular hypoplasia
 - Mandibular micrognathia
 - Broad mandibular arch
 - Narrow maxillary alveolar arch
- Malocclusion and bite abnormalities
- High arched and narrow palate
- Low tongue position
- Prominent lateral palatal ridges

Abnormal dental development

- Hypoplastic (thin) tooth enamel
- Decreased amount of dentin
- Shorter and/or bifurcated roots
- Irregular pulpal anatomy
- Reduced tooth size

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- Smaller mesial-distal dimensions of teeth
- Premature eruption and crowded teeth

Consequent Disease Pathologies

- Periodontal disease
- Tooth mobility and root resorption
- Higher incidence of decayed, missing, and filled teeth
- Growth hormones and changing craniofacial proportions

We will look at each of these conditions now.

Developmental pathologies: Abnormal oral development

Asymmetry of facial bones

The loss of the X chromosome in girls with TS affects the shape and the size of craniofacial structures. TS patients often present with retrognathia, as a result of hypoplasia or underdevelopment of the maxilla (upper jaw) and/or the mandible (lower jaw). In either case, the jaws recede significantly with respect to the forehead.

Mandibular hypoplasia (underdevelopment of the lower jaw) and micrognathia (an unusually small lower jaw) have been observed. In addition, the mandibular arch has been found to be of shorter length and broader than usual in TS patients.

In the maxilla (upper jaw) in TS patients, the alveolar arch, or top arch of teeth, is often of normal length but narrower than usual.²

The specific timing of the eruption of teeth assists the orderly development of the facial structures. A major factor in the increase of height of the maxilla is the continued apposition of bone on the free borders of the alveolar process (the projecting ridge containing the teeth and their sockets, or alveoli) as the teeth erupt.

As the maxilla descends, continued bony apposition occurs on the orbital floor (the base of the eye area), with resorption, or bone remodeling, on the nasal floor and apposition of bone on the inferior palatal surface (the top of the mouth). By alternate processes of bone deposition and modeling resorption, the orbital and nasal floors and the palatine vault move downward in a parallel fashion.³ With premature dental eruption, this pattern is compromised and proportional development is altered.

Malocclusion and bite abnormalities

Malocclusion (the misalignment of the upper and lower teeth) and bite abnormalities in TS patients are often due to a flattened cranial base angle, decreased posterior cranial base length,

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and retrognathia.⁴ Anterior and lateral open bite and lateral crossbite conditions result from differences in arch sizes in relation to each other.

High arched and narrow palate

The maxilla is generally narrow with a high, arched palate, whereas the mandible tends to be wide and micrognathic.⁵ The presence of prominent lateral palatine ridges (within the alveolar arch, at the top of the mouth) is associated with a narrower palatal width.⁶

Low tongue position

The distance of the tongue from the palate is significantly longer in TS subjects compared with controls, indicating a low tongue position in TS during palatal development. According to Gorlin et al., tongue position may result from neuromotor dysfunction of the facial muscles, a functionally restricted tongue, or from the primary palatal malformation.⁷

Prominent lateral palatal ridges

As the maxillary processes grow together toward a midline to form the hard palate, lateral ridges are molded by position of the tongue.⁸ Prominent lateral palatine ridges are related to the lack of tongue thrust into the palatal vault.⁹ A low tongue position will not flatten out these ridges, causing them to be more prominent in TS patients.

Developmental Pathologies: Abnormal dental development

Hypoplastic (thin) tooth enamel

The X chromosome regulates enamel apposition. Both X chromosomes in normal females are active in amelogenesis,¹⁰ the formation of enamel on the teeth. Research shows that the aberration of the X chromosome in TS patients most likely affects the excretion of amelogenin, the proteins responsible for enamel development, so that teeth often present enamel-related defects, such as reduced crown size and enamel hypoplasia (underdevelopment).¹¹ Teeth with thin or hypoplastic enamel are vulnerable to caries (tooth decay), especially in the presence of plaque.¹² There is also a greater potential for tooth sensitivity.

Decreased amount of dentin

The Y chromosome influences both dentin and enamel growth. The relative reduction in dentin and the estimated reduced mesial-distal width of the tooth germ in TS females indicates that their tooth development is affected at an early stage of morphogenesis.¹³

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Shorter and/or bifurcated roots

TS patients present relatively short dental roots on the incisors, canines and premolars.¹⁴ According to Lahdesmaki et al., “Premolars and lower first molars of TS females show an increased number of root components in dissimilar variants and a significantly higher frequency of two-rooted mandibular premolars than normal females.”¹⁵

Irregular pulpal anatomy

Taurodontism is an extension of the tooth body and pulp chamber in which the furcation, or branching, of the tooth roots takes place later than in a normal molar. It appears in TS females as in a normal population, a frequency of 0.5% to 5%. Exceptionally, the pulp chamber of the mandibular (lower) second premolar is often elongated in TS females.¹⁶

Reduced tooth size

All permanent teeth in TS females are reduced in size.¹⁷ A study by Zilberman et al. shows that in permanent first and second molars of TS females, crown width and the dimensions of tooth components were less than those of normal females and males, with reduction in size affecting first molars more than second molars.¹⁸ Hypoplasia and reduced dentinal formation cause reduced tooth dimensions. Incisor asymmetries and shape aberrations have also been observed in TS patients.¹⁹

Smaller mesial-distal dimensions of teeth

The mesial-distal dimensions of the permanent tooth crowns of TS females are significantly smaller than those of control females, mainly due to a thinner enamel layer,²⁰ and particularly for the lower first permanent molar.²¹

Premature eruption and crowded teeth

Unlike skeletal development, which tends to be delayed by 2+ years in TS females, dental development tends to be early. Therefore, secondary teeth may erupt prematurely.²² The first permanent molars appear between 1-1/2 and 4 years of age, rather than the typical 5-6 years of age.²³

Crowding may occur as the dentition erupts into incompletely developed jaws with shorter arch length. Alternatively, teeth may also be widely-spaced if smaller teeth erupt into jaws of normal dimension.

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Consequent Disease Pathologies

Periodontal disease

Periodontal disease may develop as a consequence of dental crowding, malocclusion with consequent food impaction, and/or reduced at-home care if limited motor skills are present.

Tooth mobility and root resorption

Loss of alveolar bone along with occlusal trauma precipitated by malocclusion may result in increased tooth mobility.

TS females are also at greater risk for root resorption, which can lead to tooth loss, especially during orthodontic treatment.²⁴

Higher incidence of decayed, missing, and filled teeth

In studies done by Capitaneanu et al., the decayed, missing, and filled permanent surfaces index for teeth was statistically higher in TS patients versus control subjects. Orthodontic anomalies were also found to be more frequent and more severe in TS patients.²⁵

Growth hormones and changing craniofacial proportions

In the human body, the one system that does not follow usual lines of development is the dentition, which, according to Kraus, “has an embryonic period that includes the entire prenatal span of time as well as the first 16 or 17 years of postnatal life ... terminating with the final calcification of the crown.”²⁶

Because tooth development continues into the teenage years, dental professionals must pay attention to whether their TS patients are undergoing treatment with growth hormone replacement therapy, as this can alter craniofacial proportions and lead to further orthodontic concerns.²⁷ The TS patient being treated with growth hormone should be assessed early for orthodontic treatment due to her unique challenges: early eruption of permanent teeth, treatment timed around major differences in growth, and differences between her chronological and skeletal ages.²⁸

Solutions

Dental care is vitally important to the overall health of any child. The American Academy of Pediatrics (AAP) recommends that children be generally seen by a dentist no later than 12 months of age if possible.²⁹

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In TS patients, high caries (tooth decay) index values reflect the need for earlier preventive measures.³⁰ The AAP recommendation for children with risk factors for caries likely to be seen in TS patients should be seen by a dentist within six months of the eruption of the first tooth or 12 months of age, whichever comes first.³¹

Some TS patients may have difficulty brushing and/or flossing in the event of limited fine motor skills. Aids such as electric toothbrushes; small, soft-bristle manual toothbrushes; end-tuft brushes for crowded areas; tongue scrapers; and floss holders can be helpful to these patients.

TS patients may also benefit from use of prescription-strength fluoride toothpaste and fluoride rinses. Rinses, especially, may be easier to use than pastes if the patient has limited dexterity.

Because TS patients typically present underdeveloped enamel, thin enamel and/or enamel defects, diet counseling for reduction of caries is recommended once a patient reaches an age at which she understands and can be responsible for her own dental health. Keeping a record of food consumption enables the dentist or hygienist to confirm that the patient is maintaining a healthy diet and to identify specific foods (such as fermentable carbohydrates) that may contribute most to plaque and to tooth decay.³²

The presence of a small and retrognathic mandible may contribute to malocclusion and other dental abnormalities. Therefore, TS patients should receive an orthodontic examination no later than seven years of age.^{33,34} Because growth hormone treatment can alter craniofacial proportions, all TS patients treated with growth hormone should receive periodic orthodontic follow-up.³⁵

While orthodontic treatment is often prescribed for TS patients, conventional treatment methods can be difficult or impossible to execute, due to severe bite abnormalities and mandibular deficiency. A combination of orthodontics and well-timed surgery (in relation to a patient's growth and development) may deliver the most successful results.

For some TS patients, treatment is more a matter of aesthetics and self-esteem building than of functionality. A variety of treatments can be recommended based on the severity of the patient's condition and the financial resources available. Extensive solutions, such as correction of retrognathia or crowding, may require the services of an oral surgeon and/or orthodontist. There are also many affordable cosmetic options such as bonding, veneers and crowns.³⁶

According to research, a congenital heart defect (CHD) occurs in approximately 30% of patients with TS. If a cardiovascular malformation is present, the TS patient should be followed by a cardiologist in collaboration with her primary physician. Patients with a structural CHD may require pre-treatment antibiotics for subacute bacterial endocarditis before dental work or any other potentially contaminating procedure.³⁷

If a child requires sedation to complete extensive dental work, a referral to a dentist with pediatric sedation certification may be needed.

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