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Turner's syndrome is a complex medical condition characterized by oral, physical and psychological features

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The Treatment Complexities of the Turner's Syndrome Patient

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Introduction

Effective patient assessment is critical to the delivery of high-quality dental hygiene care. Patient assessment extends beyond the boundaries of the oral cavity and begins when the dental hygienist first encounters the patient in the reception area. A patient's appearance, stature, gait and carriage reveal health status, indicating wellness or abnormality. Many health conditions present with a host of oral and physical manifestations.

Turner's syndrome is a complex medical condition characterized by oral, physical and psychological features.¹ The syndrome is a chromosomal anomaly seen in approximately 1 in 3,000 live births. In 1938, Dr. Henry H. Turner, an endocrinologist, characterized the syndrome as the complete or partial absence of one X-chromosome. The human genome normally consists of 46 chromosomes:

22 pairs of autosomes plus the X and Y chromosomes. Women are born with two complete X chromosomes. Turner's patients have only 45 autosomes plus one X chromosome. The

average birth length of a Turner's syndrome baby is 47 cm and its birth weight is lower than normal newborns, 2,933 ± 467 g. Approximately 98–99% of Turner's syndrome fetuses are spontaneously aborted, and about 20% of all spontaneously aborted fetuses have Turner's syndrome.^{1,2}

Common physical characteristics of Turner's syndrome patients include short stature, webbed neck and hypoplastic pinna (Figure 1), underdeveloped or shield chest with wide-spaced nipples, and edema of the hands (Figure 2) and feet. The primary systemic health risks associated with the syndrome include osteoporosis, ischemic heart disease, hypertension and stroke.¹⁻³

FROM THE EDITOR

Welcome to the premiere issue of Oral-B *Case Studies in Dental Hygiene*, a series designed to expand the knowledge base of dental hygienists on important clinical topics. Through these case reports and discussions, we hope to share information about the many complex situations you may encounter in your practices, and to address relevant diagnostic and decision-making skills you may employ. In this issue, Valerie Carter, RDH, MS looks at a fascinating case of a young woman with Turner's syndrome. Though it is unlikely that you will encounter such a patient in your practice due to the rarity of the condition, many of the counseling, educational and treatment techniques employed will have application to the challenges you face on a daily basis.

We gratefully acknowledge Oral-B for their generous support. This series is part of their commitment to our profession, and demonstrates their desire to improve the oral health of everyone.

For those of you interested in earning continuing education credits, each issue of Oral-B *Case Studies in Dental Hygiene* has been accredited for one content hour of CEU. You will earn continuing education credit by reading the article and successfully completing the quiz provided at the end of the issue.

We welcome your participation in the Oral-B *Case Studies in Dental Hygiene* series and look forward to your comments.

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Figure 1. Webbed neck, low posterior hairline and hypoplastic pinna of a Turner's syndrome infant



Figure 2. Edema of the hand of an infant with Turner's syndrome

Oral features associated with Turner's syndrome include high arched and narrow palate, occurrence of cleft palate at a higher-than-normal frequency, significantly smaller mesio-distal dimensions of the teeth, bifurcated roots and reduced root length, particularly in the maxilla, premature tooth eruption (the first permanent molars appear between 1½ and 4 years of age, as opposed to the typical 5–6 years of

age), increased molarization of premolars and reduction of cusp height and crown size, and the presence of hypoplasia. The Turner's syndrome patient's tooth enamel is often thinner than normal, with a decreased amount of dentin present.⁴⁻⁷

The physical deformities associated with Turner's syndrome also may produce psychological effects. Teenagers and young adults with the syndrome, for example, experience a tendency for low self-esteem and depression.^{1,2}

This case describes the management of a patient diagnosed with Turner's syndrome who presented with the following oral, physical and emotional conditions: severe retrognathia; posterior open bite; crossbite on the right side; generalized enamel hypoplasia; shortened roots; thin dentin; compromised dexterity; osteoporosis; mitral valve prolapse; and low self-esteem. The dentist and dental hygienist provided this patient with comprehensive care in consultation with the patient's physician and other dental specialists.

Case Report

Background and Health History

Ms. L is a 21-year-old Caucasian female with Turner's syndrome. She currently presents at 4 feet 9 inches and weighs approximately 95 pounds. She began receiving dental care at the Baltimore College of Dental Surgery (BCDS), University of Maryland, Baltimore School of Dentistry in 1987. In 1999, Ms. L sought comprehensive care at Kernan Hospital Dental Clinic in Baltimore, Maryland.

Ms. L first presented to the dental school in 1987 when she was 5 years old. Her mother (Mrs. L) requested that Ms. L receive comprehensive care. At her initial visit, Mrs. L reported that her daughter was diagnosed with Turner's syndrome at birth, with the traditional physical features of webbed neck, low posterior hairline, edema of the hands and feet, and wide-spaced nipples. Mrs. L reported that shortly after birth, Ms. L experienced a high fever due to Salmonella poisoning, resulting in "grooves" in her teeth. No known drug or latex allergies were reported.

Between 1987 and 1992, Ms. L was brought to BCDS by her mother for examination, radiographs and preventive oral health care. In 1992, a height and weight graph was developed to track Ms. L's growth patterns over time. From 1992–1999, Ms. L was below the

norm for height and weight in her age bracket (Table I). In Ms. L's dental chart, under the category of developmental history, Mrs. L. checked a box indicating her belief that her daughter had stopped growing and did not show any signs of reaching puberty.

DATE	AGE	STATURE	WEIGHT
11/03/92	10	4 feet	52 lbs.
01/17/93	11	4 ft. 2 in.	56 lbs.
03/28/94	12	4 ft. 4 in.	63 lbs.
08/18/96	14	4 ft. 8 in.	75 lbs.
02/23/98	16	4 ft. 9 in.	92 lbs.

At the age of six, Ms. L was referred for an orthodontic consultation to address her severe retrognathia. Orthodontic intervention is frequently prescribed for Turner's syndrome patients.⁸

Persons with mandibular deficiency and Class II malocclusions, such as Ms. L's, pose orthodontic challenges. Conventional orthodontic correction of the Class II deep bite deformity, accompanied by a decreased lower facial height tendency, can be mechanically difficult, inefficient and, in many cases, impossible. Overall, the best results are obtained with a combined orthodontic-surgical approach. Early evaluation and intervention are essential when combined orthodontic-surgical approaches are indicated.^{7,8} Patient evaluation is essential to decide the optimal surgical procedure and the appropriate orthodontic-surgical sequencing.^{8,9} The rate and timing of growth in Turner's syndrome patients also influence interventions. The following orthognathic options for Ms. L's mandibular deficiency and Class II malocclusions were presented: mandibular advancement with or without genioplasty; total subapical mandibular advancement; and superior or inferior repositioning of the maxilla.⁸⁻¹⁰ Despite her daughter's condition, Mrs. L declined treatment due to financial constraints.

In 1995, at age 13, Ms. L began receiving growth hormone therapy injections to increase both her height and weight; however, these treatments were discontinued in 1997. Short roots and small facial structures are apparent in her 1997 panoramic radiograph (Figure 3). Ms. L then began estrogen replacement therapy (ERT) to achieve feminization and decrease osteoporosis risk.



Figure 3. Panoramic radiograph of Ms. L (demonstrating small cranial features and shortened roots)

In June 1999, Ms. L discontinued her estrogen therapy. She took no medications until August 2001 when it was documented that she was prescribed Premarin® and Provera®. Premarin is an estrogen derivative used to treat hypogonadism

and to prevent osteoporosis.¹¹ Provera was prescribed for Ms. L's endocrine disorder and used to treat abnormal uterine bleeding resulting from a hormonal imbalance.¹¹ In early 2002, Ms. L discontinued all medications due to adverse reactions. However, in December 2002, Ms. L resumed taking Provera and Climara. Climara is an estrogen derivative used to treat female hypogonadism and prevent osteoporosis.¹¹

Ms. L's parents are alive and well. Her only sibling, a sister, is alive and free of any health concerns. Ms. L's great aunt had tuberculosis and is deceased. Her grandparents, both paternal and maternal, are alive. Socially, Ms. L does not smoke or drink alcohol, and reports that she follows a nutritious diet. Ms. L exercises regularly, is a full-time student, and is employed part-time.

Clinical Data

At her first visit to Kernan Hospital Dental Clinic, Ms. L reported having Turner’s syndrome associated with mitral valve prolapse and osteoporosis. A medical consultation regarding the patient’s need for prophylactic antibiotics was required prior to initiating any oral procedures that could induce gingival bleeding; therefore, no invasive treatment was implemented at the time. Services rendered included a medical/dental history, measurement and recording of vital signs (blood pressure and pulse), extra- and intra-oral examinations and intra-oral photography. Diet counseling was also initiated. The extra-oral examination revealed a webbed neck, low posterior hairline, broad shielded chest and a severe retrognathic profile. Intra-orally, high vaulted and narrow palate, small tooth crowns, thin enamel, enamel defects and generalized hypoplasia were documented. Bilateral linea alba and white-coated tongue were also observed.

Intra-oral photographs were taken to document Ms. L’s unique oral conditions, but Mrs. L would not allow exposure of any extra-oral photographs. Diet counseling was indicated for caries control due to the enamel hypoplasia, thin enamel and other enamel defects observed during the intra-oral examination. The dental hygienist gave Ms. L a three-day diet record form to document all foods and drinks consumed, with their amounts and times of consumption listed. Ms. L’s mother signed a form granting the physician permission to release medical information to the dentist, and a second appointment was

scheduled. Prior to Ms. L’s second appointment, the medical consultation form was returned, and the need for antibiotic prophylaxis prior to all dental procedures was noted. No other precautions were indicated.

At her second dental appointment, Ms. L reported taking 2 grams of amoxicillin one hour prior to the dental appointment, as prescribed. Vital signs were measured and recorded. Since hypertension is a risk factor in Turner’s syndrome patients,^{1,3} Ms. L’s blood pressure was monitored closely at each visit. All vital signs were within normal limits.

A panoramic radiograph and four bitewing radiographs were exposed. Ms. L’s extremely high and narrow palate with crowding made it difficult to expose the bitewing radiographs. Special care was taken with the placement of the films so that the gag reflex was not triggered, and diagnostically sound films were obtained. Panoramic radiographic evaluation, as in 1997, revealed short roots and small facial structures. There was no evidence of osteoporosis radiographically. Evaluation of the bitewing radiographs revealed no significant findings.

A hard tissue examination was performed. All teeth were present except third molars, which an oral surgeon extracted due to crowding when Ms. L was 16. Ms. L also presented with a posterior open bite, crossbite on the right side and a Class II malocclusion. Amalgam restorations had been placed on all first molars (#’s 3, 14, 19 and 30) and sealants were placed on second molars and all premolars. There were no restorations on the anterior teeth. Enamel defects were observed and documented in teeth #’s 4, 5, 6, 7, 10, 11, 12, 13, 20, 21, 22, 27, 28 and 29 (Figures 4 and 5). Malocclusion and a deep bite also were noted.

The accretions assessment showed generalized moderate supra- and subgingival plaque and calculus accumulations interproximally and



Figure 4. Intra-oral photograph of Ms. L’s generalized enamel hypoplasia and enamel defects



Figure 5. Maxillary occlusal intra-oral photograph of Ms. L’s generalized thin enamel

cervically, especially on the lingual surfaces of the mandibular anterior teeth. Minimal stain was present.

Ms. L’s periodontium revealed generalized marginal erythema, inflammation and bleeding, possibly due to a combination of plaque and ERT.¹²⁻¹⁴ The gingival tissue exhibited bulbous papillae and rolled margins. There was no recession, and probing depths ranged from 2–4 mm.

The patient’s oral health knowledge and self-care behaviors were assessed. Ms. L’s dental IQ was high. She understood the importance of good oral home care and had knowledge of the disease process. A score of 50% plaque-free surfaces was computed and a 45% bleeding index was obtained, indicating an uncontrolled condition.

Ms. L’s oral hygiene practices and techniques were fair to poor. She brushed one to two times a day with a soft-bristle toothbrush and a fluoride dentifrice, and used a fluoride mouth rinse once a day. She reported flossing only when she “had food in my teeth.” Manual dexterity was a concern. Limited fine motor skills, common among Turner’s syndrome patients,¹ affected her ability to perform thorough toothbrushing and flossing. Ms. L demonstrated a vigorous scrub technique, with the bristles pointed perpendicularly to the tooth surface. She flossed inadequately and held the floss at both ends, placing it through both sides of her mouth and “shimmying” it between her teeth. Ms. L’s low self-esteem was evident when she pointed to the enamel defects, hypoplasia and her profile, and became upset. Her low self-esteem and low motivation were significant barriers to improved oral health.

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Dental Diagnosis and Dental Hygiene Treatment Plan

The patient’s periodontal condition was diagnosed as Class I, gingivitis. No caries were detected clinically or radiographically. The patient’s occlusal classification was Class II with deep-bite, posterior open bite and crossbite on the right side. As in 1993, orthodontic treatment was recommended for correction of severe retrognathia and crowding.

A dental hygiene treatment plan was developed. The goals were to modify the patient’s oral health behaviors by targeting her low self-image, supplying her with a resource list of syndrome support systems, and by introducing specific oral hygiene aids to compensate for limited dexterity, to discuss orthodontic and aesthetic interventions, to hand scale by quadrant, to selectively polish teeth not affected by enamel hypoplasia or defects, to apply fluoride and recommend 1.1% neutral sodium fluoride gel for home use, to polish amalgams to reduce the potential for plaque retention and occlusal caries, and to institute a three-month recall interval for continued monitoring of systemic and oral health status.

Limited fine motor skills affected her ability to perform thorough toothbrushing and flossing

Implementation and Outcomes of Treatment

Education and communication were key focuses of the implementation phase of dental hygiene treatment. Since manual dexterity and malocclusion compromised Ms. L’s homed care, an electric toothbrush was recommended. Ms. L was eager and seemed motivated to try the new brush. She was asked to purchase it by her next visit, so that her technique could be observed and assessed. In case she did not purchase an electric toothbrush, manual toothbrushing was addressed and modified to incorporate sulcular brushing, particularly on the linguals of the mandibular anteriors. A small soft-bristle brush was recommended and an end-tuft brush was advised for crowded areas. Proper tongue brushing and the use of a tongue scraper were demonstrated. Due to Ms. L’s limited manual dexterity and ineffective technique, a floss holder was introduced to aid in interproximal plaque control.

Education and communication were the key focuses of the implementation phase

Patient education continued with a discussion of Ms. L’s generalized enamel hypoplasia and thin enamel and their vulnerability to caries, especially in the presence of plaque. Both the

dentist and hygienist felt that a stronger fluoride was indicated, based on the hard tissue findings and past history of estrogen deficiency. The hormone estrogen is believed to control salivary peroxidases. A decrease in the concentration of the circulating estrogens alone may cause an impaired biosynthesis of these enzymes, resulting in a compromised defense against oral bacteria.^{13,14} Due to Ms. L’s inconsistent use of ERT, a stronger fluoride seemed an appropriate preventive measure. The fluoride rinse, which contains 0.05% sodium fluoride and has a fluoride ion concentration of 0.0226%, was substituted with a 1.1% neutral sodium fluoride concentration which is more effective.¹⁵ Ms. L agreed to use the prescription fluoride toothpaste and felt it would be easier than rinsing, since it substituted for routine brushing behaviors.

During home care instructions, the patient stated, “no matter how well I take care of my teeth, they will still be ugly like me.” Communication at this point required caring and compassion. Ms. L’s emotional state presented an opportunity to build dental hygienist/patient rapport. When Mrs. L decided to leave the treatment room and wait in the reception area, an intimate discussion of Ms. L’s school and personal feelings ensued. The patient revealed her depressed feelings about her “looks” and how she was teased and stared at by others. The dental hygienist sought to build Ms. L’s confidence and self-esteem. She complimented Ms. L on her accomplishments and positive traits, and explained that improved oral hygiene could enhance her appearance and well-being.

A major concern related to the patient’s self-image was her profile. In complex Class II malocclusion cases, combined surgical-orthodontic approaches can provide increased treatment efficiency, long-term stability and optimal esthetic results,^{8,9} so both an oral surgeon and orthodontist were consulted. They presented treatment options for the correction of severe retrognathia and crowding that were similar to those proposed in 1993. The general dentist and hygienist also proposed less costly cosmetic options such as bonding, veneers and crowns to aid in the esthetics of Ms. L’s maxillary anterior teeth. The dental hygienist met with Mrs. L to discuss her daughter’s self-esteem issues and to provide further information regarding treatment options, but Mrs. L declined all suggested treatment due to finances.

At the next appointment, Ms. L accompanied the dental hygienist

to the treatment room without her mother. Despite the setback of not proceeding with surgical or cosmetic interventions, Ms. L. seemed upbeat, initiating a conversation regarding her school and work. After disclosing, Ms. L’s plaque score showed an improvement, from 50% to 75% plaque free. Scores were charted and served as a motivational tool. Ms. L brought the electric toothbrush to the appointment and demonstrated her technique. Slight brushing modifications were made with angulation and pressure. Ms. L was extremely happy that her mother had purchased the brush and she, in return, was committed to using it.

Ms. L presented her three-day diet diary form and stated that she felt more responsible for her own health by completing it. Fermentable carbohydrates were circled, and a discussion regarding quantity and frequency followed. Overall, Ms. L’s diet was healthy and well-balanced.

Ms. L stated that she pre-medicated with amoxicillin one hour prior to her dental appointment, so hand scaling began. Ultrasonic scaling was contraindicated due to the enamel defects,¹⁶ the brittle nature of Ms. L’s teeth and the potential for sensitivity due to her thin enamel.¹⁶ The mandibular right and maxillary right quadrants were scaled to completion with generalized moderate bleeding. Ms. L was comfortable and compliant throughout all procedures. She observed the results and was pleased at the appearance of her “clean” teeth. Ms. L appeared motivated to improve her oral health status.

The patient was reappointed for completion of the maxillary and mandibular left quadrants. She confirmed taking her pre-medication and demonstrated a new confidence previously unseen. A 90% plaque free score was charted. Home care instructions were reviewed and a brushing angulation modification was made so that Ms. L could better access the linguals of the mandibular anterior teeth. The maxillary and mandibular left quadrants were hand scaled to completion and amalgams were polished. Coronal polishing is contraindicated on teeth with enamel hypoplasia,¹⁶ so selective polishing was performed and a fluoride treatment was administered with post-care instructions.

Over the course of treatment, Ms. L’s home care improved from an initial score of 50% plaque free to a 90% plaque free score at the re-evaluation appointment. Gingival tissue response to scaling was evident. The gingival tissue became more firm, pink and healthy, filling the interdental spaces. The marginal gingivitis had resolved with Ms. L visualizing the difference between health and disease.

Ms. L’s confidence was improved despite the financial constraints that precluded orthognathic and aesthetic interventions. Her decision to take control of her oral health was evident. The fact that her mother was attentive by purchasing her electric toothbrush and prescription fluoride had a positive influence on Ms. L’s oral health and self-esteem. Ms. L also felt responsible for her oral health by completing the diet diary and performing her prescribed oral hygiene regimen.

Her mother’s purchase of an electric toothbrush and prescription fluoride had a positive impact on Ms. L’s oral health and self-esteem

She also stated that when she was more financially stable, she would seek orthodontic care on her own. She was placed on a three-month recall for preventive care. Ms. L’s prognosis is very good.

Discussion

The key discussion points related to this case include the physical manifestations and recognition of the Turner’s syndrome patient,



For everything you do

- [scaling and polishing]
- [perio debridement]
- [fluoride treatments]
- [hand-holding]
- [words of encouragement]
- [words of wisdom]
- [smiling even though your
back is killing you]

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precisely

the importance of physical assessment as related to oral health care, treatment interventions available to the patient, and the need for oral health care providers to address the psychological aspects of patient care. These topic areas are discussed below.

Turner's syndrome patients present with a broad range of oral and physical characteristics. These features may vary in severity; if pronounced at birth, the Turner's syndrome patient can be recognized readily. If diagnosis is not made at birth, then Turner's syndrome is suspected at puberty when sexual maturation fails to occur at the expected age.^{1,2}

A consistent feature in Turner's syndrome is small stature.^{1,2} The rate of growth for the patient may be normal during the first 3 to 5 years of life, but after age 5 it may start to decelerate, resulting in a significantly short stature.^{1,2} Oral health care providers may play a role in the identification of Turner's syndrome patients, especially in a pediatric practice where monitoring height, weight and growth are critical.

In Ms. L's case, both growth hormone replacement therapies and estrogen replacement therapies were prescribed. Treatment with recombinant human growth hormone increases height and stature in most, but not all children.^{1,12,13,17} Some believe growth hormone treatments should be started early in childhood, but many wait until there is evidence of deceleration of growth on a specific Turner's syndrome growth curve.^{1,2,12} Ms. L began growth hormones at age 13.

Replacement therapy with estrogens is indicated in the Turner's syndrome patient to achieve feminization, but there is no definitive age at which to start treatment.^{1,13} Psychological preparedness of the patient must be considered when initiating therapy. The improvement in growth achieved by the girls treated with growth hormones allows initiation of estrogen replacement at the age of 12 or 13.^{1,13} Greater gains in height have been seen when estrogen replacement is initiated at 13 or 14 years of age.^{1,12,13,17} Ms. L began her estrogen replacement therapy at age 15. Her inconsistent use of growth hormones and ERT may partially explain her shortened stature.

Turner's syndrome is associated with many medical complications. Table II presents features of Turner's syndrome (45,X). Medical complications include non-stenotic bicuspid aortic valves that occur in about one-third to one-half of patients. In later life, bicuspid aortic valve disease can progress to dilatation of the aortic root. Less frequently occurring defects include aortic coarctation, aortic stenosis, mitral valve prolapse, and anomalous pulmonary venous drainage. The webbed neck in patients with or without syndromes is associated with both flow- and non-flow-related heart defects.¹⁻³ Since Ms. L already presents with mitral valve prolapse and webbed neck, she should be monitored for future heart disease. Evidence also supports the important role of oral health status in the prevention of bacterial endocarditis (BE) of dental origin. When treating patients at risk for BE, oral health professionals should emphasize the importance of oral health care.¹⁸

Seventy-five percent of Turner's syndrome patients develop recurrent bilateral otitis media and sensorineural hearing deficits with frequency increasing with age.^{1,2} Although Ms. L did not present with hearing deficits, this feature of the Turner's syndrome patient could impact oral health care, requiring the dental hygienist to communicate in an audible volume without embarrassing the patient. Visual,

Table II
Features of 45, X Turner's Syndrome¹

CLINICAL ABNORMALITIES	PERCENT OF FINDINGS	CLINICALLY OBSERVED
Webbing of neck	50%	X
Edema of dorsa of hands and feet	20%	X
Low posterior hairline	80%	X
Small mandible	70+%	X
High arched palate	80+%	X
Prominent ears	80%	X
Epicanthal folds	40%	X
Broad chest	80+%	X
Cubitus valgus	70+%	X
Hyperconvex fingernails	70%	X
Bone dysplasia	50+%	X
Horse-shoe kidney/double or cleft renal pelvis	60+%	
Cardiac defects:	30+%	
Coarctation of aorta	20%	
Mitral valve prolapse	46%	
Scoliosis	10%	X
Hearing impairment	75+%	X
Hypertension		X

as well as verbal directions would be indicated for patient education.

Turner's syndrome patients with developmental problems in fine motor skills also may have limited dexterity¹ and experience problems performing oral self-care procedures, as was evident with Ms. L. Dental hygienists, therefore, need to be creative in suggesting techniques for tooth brushing and flossing.

Dental diseases and disorders can interfere with social

roles at any age. These interferences may damage self-image and alter one's ability to sustain and build social relationships.¹⁹ Teenagers and young adults with Turner's syndrome experience a tendency for low self-esteem and depression.^{1,2,14,19,20} Attitude and mental state play major roles in patients' willingness to accept responsibility for their own oral health care.¹⁹ The dental hygienist's treatment plan needed to be modified to address Ms. L's low self-esteem and emotional needs. Had Ms. L's self-esteem not improved, consultation with the patient's physician and possible referral for psychological assistance may have been necessary.

Ms. L, like many Turner's syndrome patients, presented with thin enamel and generalized enamel hypoplasia,¹ a defect that occurs as a result of a disturbance in the formation of the organic enamel matrix.¹⁶ Ms. L's medical history disclosed an illness with high fever in childhood which may have compounded pre-existing hypoplasia and enamel defects associated with Turner's syndrome (Figures 4 and 5).

Ms. L also presented with severe retrognathia, which affected her profile. Patients with Turner's syndrome, like Ms. L, may have a cranial base that is short, and a facial profile that is retrognathic, with the mandible being short and the maxilla being of normal length.²¹⁻²⁵ These distinctive characteristics, as well as the rate and timing of growth, significantly influence orthodontic treatment¹⁰ and form the wide flat-shaped facial characteristics of Turner's syndrome patients.^{21,22} The patient who is taking growth hormone replacement therapy needs to be assessed early for orthodontic treatment due to the challenges of early eruption of permanent teeth, altered treatment timing because of major differences in growth, and differences between chronological and skeletal ages.^{10,21,23-28}

Osteoporosis is a systemic health risk for the Turner's syndrome patient.¹ Oral bone loss can result from several systemic diseases, and has been linked with periodontitis, residual ridge resorption and age-related systemic osteoporosis.²⁹ Early detection of osteoporosis helps identify those at risk, and enables them to take preventive measures. Dentists and dental hygienists routinely take intra-oral radiographs so

Dental diseases and disorders can interfere with social roles at any age

they can be instrumental in early detection.²⁹ In Ms. L's panoramic radiographs, there was no evidence of osteoporosis, but with the knowledge of her syndrome, the dentist and dental hygienist should evaluate Ms. L's radiographs at each visit to monitor bone density changes.

Turner's syndrome patients are aware of their condition and its associated risks. Women with Turner's syndrome have a reduced life expectancy due to an increased risk of aortic dissection and ischemic heart disease.^{1,3} Until recently, women with Turner's syndrome did not have access to focused health care, and thus quality of life was reduced in a significant number of women.^{1,4} Ms. L is aware that she is at higher risk for disease and needs to be followed by a multidisciplinary team to improve her life expectancy and reduce her morbidity.

Dental hygiene treatment for the Turner's syndrome patient offers many challenges. Holistic care is critical. Observing the patient in the reception area and treatment room, taking a complete and thorough medical history, documenting medications and assessing vital signs allow for comprehensive care and the formulation of individualized treatment plans. Clinical manifestations, medications and the patient's attitude pose additional challenges. Oral health professionals may be instrumental in the initial identification of the syndrome and in recognizing subsequent emotional problems, cardiac conditions and osteoporosis. Triage with dental and medical specialists, often indicated when treating Turner's syndrome patients, adds an exciting dimension to patient care and promotes the delivery of high quality dental hygiene services.

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CE QUIZ

- All of the following are common characteristics of the Turner's syndrome patient except
 - webbed neck.
 - hypertension.
 - cardiac defect.
 - low posterior hairline.
 - butterfly rash over nose.
- Turner's syndrome is a chromosomal anomaly seen in approximately how many live births?
 - 1 in 2,500
 - 1 in 3,000
 - 1 in 2,000
 - 1 in 5,000
- A medical consultation with the patient's physician is recommended for the Turner's syndrome patient for what condition?
 - Webbed neck
 - Bone dysplasia
 - Cardiac defects
 - Scoliosis
- Oral manifestations occurring in the Turner's syndrome patient may include all of the following except
 - premature tooth eruption.
 - high arched palate.
 - smaller medio-distal tooth dimensions.
 - thinner enamel.
 - mulberry molars.
- Approximately 98 to 99% of Turner's syndrome fetuses are spontaneously aborted?
 - True
 - False
- Complications seen in the Turner's syndrome patient may include all of the following except
 - developmental delay in fine motor skills.
 - tendency for low self-esteem.
 - renal malformations.
 - hepatitis.
- Systemic estrogens are used to achieve feminization in the Turner's syndrome patient. Which of the following is not an estrogen?
 - Premarin
 - Provera
 - Climara
 - Estratab

- 8. Optimal orthodontic treatment of the Class II deep-bite malocclusion for Turner's syndrome patients include all the following **except**
 - a. early identification.
 - b. an orthodontic-surgical approach.
 - c. a specific timing sequence.
 - d. extractions of second bicuspid.
- 9. All of the following factors caused modifications in the case study patient's dental hygiene treatment plan **except**
 - a. decreased manual dexterity.
 - b. mitral valve prolapse.
 - c. financial issues.
 - d. uncooperative behavior.
- 10. What is the chromosomal abnormality associated with Turner's syndrome?
 - a. XX
 - b. XY
 - c. X
 - d. XXY
- 11. The rate of growth for the Turner's syndrome patient may be normal during _____ years of life.
 - a. the 1st to 2nd
 - b. the 3rd to 5th
 - c. the 7th to 10th
 - d. the 11th to 13th
- 12. What dental specialist could be instrumental in diagnosing Turner's syndrome?
 - a. Oral surgeon
 - b. Periodontist
 - c. Endodontist
 - d. Pedodontist
- 13. Which of the following conditions contraindicated coronal polishing in the dental hygiene treatment plan of the Turner's syndrome patient in this case?
 - a. Small tooth crowns
 - b. Dentinogenesis imperfecta
 - c. Enamel hypoplasia
 - d. High arched palate
- 14. Turner's syndrome patients present challenges to the orthodontist due to all of the following **except**
 - a. early eruption of permanent teeth.
 - b. growth hormone therapy.
 - c. macroglossia.
 - d. differences in chronological and skeletal ages.
- 15. The main systemic health risks associated with Turner's syndrome includes all of the following **except**
 - a. ischemic heart disease.
 - b. stroke.
 - c. hypertension.
 - d. edema of hands and feet.

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