DOWN SYNDROME

A REVIEW FOR DENTAL PROFESSIONALS

Purpose of this Module

The information presented in this module is intended to provide dental providers with an in-depth review of the medical and dental implications of Down syndrome and suggestions for dental treatment and preventive regimens based upon this detailed knowledge.

Learning Objectives

After reviewing the enclosed information the participant shall be able to:

1. Describe the three genotypes of Down syndrome.
2. Discuss the prevalence of Down syndrome, particularly related to maternal age.
3. Describe the characteristics of mid-face hypoplasia common to individuals with Down syndrome.
4. Discuss the relationship of Down syndrome to Alzheimer's disease
5. Discuss the relationship of Down syndrome to atlantoaxial instability.
6. Discuss the issues and impact of congenital heart disease among persons with Down syndrome.
7. Describe the impact of upper respiratory infections on the treatment of persons with Down syndrome.
8. List and discuss additional medical conditions common to persons with Down syndrome.
9. Discuss in detail the periodontal and oral hygiene issues involved with Down syndrome.
10. Describe malocclusion problems common to persons with Down syndrome.
11. Describe treatment approaches to the periodontal problems associated with Down syndrome.
12. Discuss the issues impacting prosthodontic services for individuals with Down syndrome.

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INTRODUCTION

Dental providers who treat patients with Down syndrome should be familiar with the medical and dental implications of this condition. Although generalizations are helpful to better understand the impact of Down syndrome on the provision of dental care, it is important to also be aware of the wide variance in expression of the various aspects of this genetic syndrome.

Definition and Etiology

Down syndrome was first described by Dr. Langdon Down in 1865. At that time, the diagnosis of the syndrome was based solely on physical findings. It was only in 1956 that the normal complement of 46 human chromosomes was determined and it was not until 1959 that it was demonstrated that Down syndrome was associated with an extra chromosome of the twenty first group, for a total of 47 chromosomes. The etiology of Down syndrome relates most often to nondisjunction of chromosome 21 during oogenesis, thus an extra chromosome 21 may be provided to the offspring by the mother. While nondisjunction of chromosome 21 does occur in spermatogenesis and can result in Down syndrome, spermatogenesis often does not continue when such nondisjunction occurs. Thus, less than 10% of cases of Down syndrome are attributable to paternal etiology.

There are three different causes of Down syndrome. Although it is generally thought that there are no clinical differences in the various genotypes, research into genotype-phenotype correlation is ongoing.

(1) Trisomy 21 (94%): The extra copy of chromosome 21 (three instead of the usual two) produces a complement of 47 chromosomes.
(2) Translocation (5%): A segment of chromosome 21 is found attached to another chromosome (usually #14, thus referred to as a 14/21 translocation). These individuals have a complement of 46 chromosomes.
(3) Mosaicism (1%): Nondisjunction occurs during early cell division after conception. Therefore, some cells have a normal complement of 46 chromosomes and other cells have 47 chromosomes (with an extra 21 chromosome).

Prevalence and Life Expectancy

The overall population prevalence is estimated to be over 250,000 individuals with Down syndrome living in the United States in 2008. Over time, this prevalence has increased along with the increase in childbearing age. The birth prevalence of Down syndrome is approximately one in 700 live births and increases with the age of the mother. The birth prevalence for a maternal age of 20 is 1 in 2300 live births while for a maternal age of 34-39 it is 1 in 280 live births. For a maternal age of 40-44 the birth prevalence is 1 in 130 live births and for a maternal age of 46 it is 1 in 65 live births. Adults with Down syndrome are living longer with life expectancy increasing dramatically over the past century (9 years in 1929, 30 years in 1980, and 55 years in the early 2000s).

Association with Intellectual Disability

Although there are some persons with Down syndrome with an IQ of 70 or over, nearly all persons with this condition have an intellectual disability. All ranges of intellectual disability can occur but most have mild or moderate intellectual disabilities (42% mild, 26% moderate, 15% severe, 17% profound). Additional information about the care of dental patients with intellectual disabilities is presented in Module 2.

FACIAL CHARACTERISTICS

Midface hypoplasia, appearing as a hypoplastic middle third of the face, is a cardinal characteristic of persons with Down syndrome. Additionally, a reduced nasal protrusion (a flat, broad bridge of the nose) has been reported in 59-78% of those with Down syndrome. Ear malformations, including congenital lop ear, low-set ears and ears with a flat or absent helix have been reported in 54% of cases. Eye abnormalities are common. Epicanthal folds with slanting almond-shaped eyes (narrow palpebral tissue slanting toward the midline) are reported in 78% of this population. Strabismus (cross eyes) is reported in 14-54% and nystagmus (constant involuntary cyclical movement of the eyeballs) and refractive errors are also common. The majority of persons with Down syndrome exhibit brachycephaly (a broad, short head). Lack of supraorbital ridges and hypotelerism (secondary to hypoplasia of the central face) are common findings. Absence of frontal sinuses and absent or reduced maxillary sinuses have been reported. Nasal septum or nasal conchae deviations are often observed which can produce a partially obstructed or narrow air passage and can contribute to mouth breathing.

MEDICAL CONDITIONS

Heart Disease

Approximately 50% of individuals with Down syndrome have congenital heart disease. For those with Down syndrome, the most common congenital heart disease in the United States is atrioventricular septal defect (AVSD, also known as atrioventricular canal defect), which occurs in 45-60% of cases. Other common congenital heart conditions in this population include ventricular septal defect (VSD) at 32-35%, atrial septal defect (ASD) at 1-8%, patent ductus arteriosis (PDA) at 7% and tetralogy of Fallot at 4-6%. The rate of these defects in individuals with Down syndrome varies across countries with researchers citing possible environmental and genetic influences as the reason for this variance.

There is also a risk for valvular disease in this population. A 40% prevalence of mitral valve prolapse (MVP) has been reported in persons with Down syndrome. The diagnosis of MVP and aortic regurgitation (AR) is much higher when echocardiograms are used. In one study, a medical history showed 6% of subjects with Down syndrome (versus 2% of controls) had aortic regurgitation and 14% of the subjects with Down syndrome (versus 4% of the controls) had mitral valve prolapse. When echocardiograms were obtained on the subjects with Down syndrome the prevalence of AR rose to 11% and that of MVP rose to 57%. A connective tissue abnormality (collagen defect) may be a possible explanation of the high incidence of MVP in this population.

The need for antibiotic prophylaxis in these patients will depend on current American Heart Association (AHA) guidelines. For patients with Down syndrome, consultation with a cardiologist is indicated when the medical history provided is unclear as to the cardiac status and history or whenever further

clarification is needed. The 2007 AHA guidelines recommend endocarditis prophylaxis for those with prosthetic cardiac valves (or prosthetic material used for cardiac valve repair), those with previous cases of infective endocarditis, those who received a cardiac transplant and developed valvulopathy, and in the following cases of congenital heart disease:

- Unrepaired cyanotic congenital heart disease, including palliative shunts and conduits
- Completely repaired congenital heart defect, with prosthetic material or device, within the first 6 months after the procedure
- Repaired congenital heart disease with residual defects at the site or adjacent to the site of a prosthetic patch or prosthetic device (which inhibit endothelialization)

It would be prudent to review current AHA recommendations on prophylaxis for infective endocarditis prior to proceeding with dental treatment for this population.

Immunologic Response

Children and adults with Down syndrome have increased risk for infection compared to the general population. Evidence of immune system differences are numerous and include a smaller thymus, decreased number of leukocytes (including B-cells and T-cells), impaired chemotaxis of polymorphonuclear lymphocytes (PMNs) and impaired T-cell function. The higher prevalence of upper respiratory tract infections (URIs) and ear infections seen in persons with Down syndrome is thought to be due to the impaired immunologic response to infectious or inflammatory diseases. Recurrent otitis media is a common finding. There is a higher risk of pneumonia, especially in young children. Prior to the age of antibiotics, most of these individuals died at an early age from pneumonia. The increase in URIs contributes to mouth breathing and speech problems and along with nasal septal deviations, may complicate the use of general anesthesia.

While leukocytes are generally reduced in numbers for individuals with Down syndrome, the prevalence of leukemia, an unrestrained growth of leukocytes, has been reported as 15-50 times higher in this population. The presentation of leukemia differs for individuals with Down syndrome compared to those without Down syndrome. The myeloid-type of leukemia originates from myeloid blood cell precursors, the cells that eventually form granulocytes as well as red blood cells and platelets. This condition, referred to as acute myeloid leukemia (AML) when presenting in patients without Down syndrome, is sometimes referred to as Down syndrome myeloid leukemia (DS ML) due to its varying clinical features. DS ML often presents with an early phase of myelodysplasia and responds well to chemotherapy (cure rate of 80%) due to the differences in the myeloid leukemia cells in those with Down syndrome compared to those without Down syndrome. For acute lymphoblastic leukemia (ALL) in patients with Down syndrome the leukemic cell-line develops almost exclusively from B-cell precursors. Patients with Down syndrome do not tolerate traditional chemotherapeutics used for ALL, such as methotrexate or anthracyclines. Historically, the prognosis was generally worse for those with Down syndrome ALL than for those with non-Down syndrome ALL; however, some modified treatment regimens have shown the ability to improve the ALL cure rate for this population to match the ALL cure rate for those without Down syndrome.

Despite the increased risk of leukemia, persons with Down syndrome are at a decreased risk of solid tumors.

Dementia

Approximately half of all adults with Down syndrome have clinical signs of cognitive deterioration occurring prior to 50 years of age. Despite evidence that many of these individuals live long lives with no clinical signs of dementia, many studies show that virtually 100% of persons with Down syndrome over 35 years old develop neurological changes associated with Alzheimer's disease, as evidenced by post-mortem findings. The discrepancies between neurological and clinical findings may be explained by the evidence that although the neurological changes seen in persons with Down syndrome and Alzheimer's disease are similar, the regional distribution within the brain is different. As individuals with Down syndrome are living longer than in previous generations and the population prevalence is increasing, the prevalence of Down syndrome-related dementia may be increasing as well.

Endocrine Problems

Children and adults with Down syndrome have an increased risk of thyroid problems compared to those without Down syndrome. The rate of congenital hypothyroidism among infants with Down syndrome is 1.5 to 6.1%, a risk that is 28 to 35 times higher than the general population. Newborn screening and treatment may reduce the growth problems and intellectual impairment associated with congenital hypothyroidism.

Subclinical hypothyroidism, seen as a mild elevation in thyroid stimulating hormone (TSH), is present in 25-60% of those with Down syndrome. It can be difficult to determine if individuals with Down syndrome have symptoms of hypothyroidism. Such symptoms are often noted in those with Down syndrome regardless of thyroid function. There is considerable disagreement in the literature on the treatment of subclinical hypothyroidism in patients with Down syndrome, with many suggesting yearly screening and early treatment. Others note that subclinical hypothyroidism in this population is often self-limiting and recommend avoiding unnecessary interventions and testing. Some researchers suggest that tests of thyroid function in individuals with Down syndrome not be compared to the general population. The reason cited is that those with Down syndrome have a different normal range for these values.

Autoimmune hypothyroidism is seen in 7.5-34% of children and adults with Down syndrome that are 8 years of age or older. As opposed to the general population where a higher proportion of females are affected, autoimmune hypothyroidism appears to affect both males and females equally in this population. Graves disease, a type of hyperthyroidism, is seen with increased frequency in this population. However, the risk is still small, with studies reporting 0-3% prevalence in those with Down syndrome.

In addition to thyroid dysfunction, diabetes is another endocrine condition with increased frequency in the population with Down syndrome. The rate is three times higher than in the general population for both type 1 and type 2 diabetes mellitus (DM). The type 1 DM onset is also somewhat earlier for those with Down syndrome (8.2 versus 8.4 years). Celiac disease, often seen in combination with endocrine disorders, is also elevated in this population (20%).

Breathing and Airway

There is an increased risk of obstructive sleep apnea in this population due to anatomical features, with 50-80% of the population affected. Aspiration of liquids, foods, saliva or gastric secretions is another common airway problem. Individuals with Down syndrome are at risk of aspiration due to hypotonia that

often results in dysphagia (58%) and gastroesophageal reflux disease (59%). Additional airway and pulmonary problems seen with increased frequency in the population with Down syndrome include upper respiratory tract infections as previously discussed, laryngomalacia, tracheabronchomalacia, tracheal bronchus, pulmonary hypertension, subpleural cysts, and subglottic stenosis.

The effects of frequent upper respiratory tract infections, obstructive sleep apnea, aspiration risk, and other airway and pulmonary problems may complicate the use of sedation in persons with Down syndrome. Special attention to patient positioning and sedation monitoring, especially with pulse oximetry, is indicated. In some cases, patients requiring sedation may require care in a hospital setting.

Atlantoaxial Instability

The prevalence of atlantoaxial instability in persons with Down syndrome has been reported as ten to twenty percent. This condition refers to an abnormal increase in mobility of the upper two cervical vertebrae (Cl/C2) due to congenital ligamentous laxity. What percentage of instances of instability leading to actual dislocation, which can lead to severe spinal cord injury, is unknown. Although some sources recommend early (5-6 years) clinical and radiographic evaluation in these individuals, others claim that instability leading to dislocation is not well founded and that radiographs are not predictive in this regard. This problem of ligamentous laxity apparently decreases with age, similar to the age-related increase in degenerative cervical arthritis which is also commonly seen in these individuals.

Being aware of the potential for atlantoaxial instability in this population is important for dental providers, especially when considering the use of medical stabilization for a patient resisting dental care or for sedation or general anesthesia when intubation is needed. Patients should be evaluated for this condition by a neurologist or neurosurgeon prior to care where significant manipulation of the head is possible (e.g., intubation for general anesthesia). For general anesthesia, these cases should be managed in a hospital setting and should be reviewed with the anesthesiologist in advance.

Ligamentous laxity can affect other parts of the body producing a hyperflexibility of all joints. This problem together with general muscle hypotonia can produce the shuffling gait often seen in persons with Down syndrome.

Other Medical Problems

- **Speech:** The speech problems commonly noted are related to the central motor deficit and degree of intellectual disability rather than a peripheral articulation problem. Delayed speech and a husky quality of voice are common findings. Speech problems may make communication difficult for some individuals with Down syndrome.

- **Hearing:** Studies show that persons with Down syndrome have a significantly higher level of hearing impairments with a prevalence of 77% reported. This hearing impairment, usually mild, is related to smaller ear canals and consequent impacted cerumen. The potential for hearing loss should be taken into account when communicating with individuals with Down syndrome.

- **Eye problems:** There is an apparent increased risk for cataracts in those with Down syndrome.

- **Other findings:** Persons with Down syndrome usually are below average in height and often have a stooping posture. Obesity and sexual underdevelopment are common. Dry, rough, scaly skin is also

common, and a single palmar crease is seen in 45% of cases.

DENTAL CONDITIONS

Periodontal Disease/Oral Hygiene

The population with Down syndrome has a higher rate of periodontitis than the general population. There are also increased rates of periodontitis compared to specific disability groups, such as those with cerebral palsy or intellectual disability. Early onset of periodontitis is a common finding (58-96%). It is possible to see alveolar bone loss in persons with Down syndrome at 6-16 years old. Additionally, the periodontal destruction occurs at a more rapid rate.

There are several proposed mechanisms for the high rate of periodontitis in this population. Some include differences in morphology and function, such as mouth breathing and small teeth and roots. The issue of ligamentous laxity/degeneration may also play a part in the destruction of the periodontal ligament. Research has revealed mixed results on bacterial profiles of periodontal pathogens, showing either similar or dissimilar bacterial profiles for those with Down syndrome compared to those without Down syndrome. The evidence shows, however, that persons with Down syndrome generally do not experience a different level of oral hygiene than other persons with intellectual disability, nor do they have increased calculus levels.

The main mechanism for increased periodontal disease is thought to be directly related to the altered immunologic response to infectious and inflammatory disease reported in this population. This defect in host response has been reported in many controlled studies showing greater inflammation and cellular response occurring with equal plaque levels. There is decreased neutrophil chemotaxis and phagocytosis. There is an increase in oxidative radicals which can cause cellular damage. There is also an increase in certain inflammatory mediators, including prostaglandins (PGE2) and leukotrienes (LTB4). There are different cytokine compositions with some cytokines that appear to be overexpressed (T-helper related cytokines IFNγ and IL4). Additionally, there are several matrix metalloproteinases (MMP2, MMP8, MMP9) that are increased for those with Down syndrome. These factors cause damage to multiple tissues, resulting in bone and attachment loss.

The clinical course in individuals with Down syndome is similar to early-onset periodontitis, except it is not isolated to a few teeth. Additionally, periodontal destruction is more rapid in this population. Several reports have indicated a high incidence of necrotizing ulcerative gingivitis (NUG) in this population. One institutional-based study showed 45% of subjects with Down syndrome compared to 4% of subjects with intellectual disability had evidence of NUG. The clinical picture of NUG in persons with Down syndrome differs from the usual symptoms in that fetid breath and exquisite pain are rarely reported. This condition historically has been associated with alterations in host response, often stress related. Oral conditions associated with an increase in NUG in this population include crowded dentition, traumatic occlusion, peg shaped anterior teeth, lack of root resorption in primary teeth, and incidence of high frenum attachment.

Caries

Individual studies from multiple countries have shown that those with Down syndrome are more likely to be caries-free compared to sibling controls and other groups with intellectual disabilities. For example, two studies found a caries-free rate of 72-78% of children with Down syndrome compared to 46-58% of their siblings. Another study found a caries-free rate of 69% of those with Down syndrome compared to 45% of those with

intellectual disability. A forth study found the caries-free rate to be higher for children with Down syndrome compared to children with and without other disabilities at all age groups. While there are some studies that show no difference in caries rates between those with Down syndrome and those without, the majority of studies demonstrate reduced caries experience in the population with Down syndrome.

Several researchers have attempted to elucidate reasons for lower caries experience in the population with Down syndrome. One active area of research on reduced caries risk in Down syndrome is altered salivary composition. While the salivary flow rates are generally lower in those with Down syndrome, there are several potential protective mechanisms of altered salivary composition that researchers have considered. Some of the proposed mechanisms studied, often with conflicting results, include changes in salivary buffering capacity, differences in salivary pH, changes in salivary electrolytes, differences in salivary proteins, and the presence of salivary IgA (both total and *Streptococcus mutans* specific). In addition, some studies have seen different levels of *S. mutans* and different *S. mutans* genotypes among those with Down syndrome compared to controls. Additional physical factors may include delayed eruption, fewer teeth (due to congenitally missing teeth), smaller teeth, and wider interdental spaces. Environmental factors might include different living conditions, dietary habits and oral hygiene habits.

Malocclusion and dental anomalies

There is an increased prevalence of malocclusion in individuals with Down syndrome, particularly Class III malocclusions. The higher incidence of Class III malocclusions is due to the underdevelopment of the midface (nasal, premaxillary and maxillary bones) not to absolute prognathism. The increased prevalence of all malocclusions has been reported as: Class III, 32-70%; Class II, 3-32%; posterior unilateral or bilateral cross bite, 71%; and open bite, 5%.

The palatal shape is small in all dimensions with a decreased arch length and arch circumference. In approximately 29% of cases the palatal vault is normal, compared to 52% in controls. In 33% of cases, the vault is furrowed, a rate similar to the control rate of 36%. A significant portion of those with Down syndrome have a shelf-like palate compared to controls (38% compared to only 12% of controls). This shelf-like pattern is believed to be due to incomplete contact between a hypotonic tongue and palate, mainly contacting on the lateral palatal walls. This pattern tends to reduce with aging due to increased lingual muscle tone.

Microdontia, especially in the mesio-distal dimension is common. The prevalence of missing permanent laterals has been reported as high as 35-43% compared with 2% in the general population. Rotated teeth (especially centrals), spaced teeth (especially lower bicuspids), peg shaped teeth (especially laterals) and more congenitally missing permanent teeth are common findings.

Additional findings include delayed eruption of permanent teeth (average 0.7 years, maximum 2 to 3 years), more impacted teeth (especially canines and premolars), more over retained primary teeth (especially primary canines and second molars), and more variable tooth morphology (including conical teeth, peg teeth, and shovel shaped maxillary incisors). There is evidence that permanent teeth in this population are shorter than average, especially in the roots, creating an unfavorable crown to root ratio. Increased taurodontism (apical displacement of root bifurcation and floor of the pulp chamber) has also been reported in persons with Down syndrome. Taurodontism, together with abnormally short roots, would reduce the extent of periodontal ligament attachment and

may contribute to tooth mobility often seen in this population.

Other Dental Problems

The prevalence of macroglossia has been reported as 11-60% in persons with Down syndrome. Whether this is due in-part to an absolute macroglossia has been debated by some investigators. There is agreement, however, on the presence of relative macroglossia due to the small palatal space and hypotonic tongue. Fissured tongue and protruding tongue due to the relative forward position of the mandible and open mouth is a common finding. Increases in bifid uvula, submucous clefts and cleft palates have been reported in this population. Mouth breathing is also a common finding.

DENTAL TREATMENT CONSIDERATIONS

The following information is derived from the literature regarding treatment options for persons with Down syndrome and suggestions presented by the authors. The management of behavioral supports for persons with intellectual disabilities is covered in Modules 2, 5 and 6.

Periodontal Disease and Oral Hygiene Problems

Early communication with patients, parents and guardians regarding the limitations of dental care in preventing tooth loss and other dental sequelae including prosthetic limitations is important. The responsibility for meticulous oral hygiene and the support and development of healthy dental behaviors by the parents and caregivers should be emphasized. Some patients may be able to effectively brush their own teeth, but most will not have the hand coordination and/or ability to follow the complex sequence of events required for effective plaque removal. Watching the patient or caregiver brush the patient's teeth may help determine any additional guidance needed. Demonstration of oral hygiene techniques may be beneficial. With high caregiver turnover, some patients may not receive consistent home care and may need continued re-introductions to home care at subsequent visits. Flossing aids may prove beneficial. Early dietary counseling can also be of benefit in preventing the nutritional problems commonly encountered in this population.

Early and Innovative Periodontal Therapy

Early and innovative approaches based upon knowledge of the host response problems may prove fruitful in addressing the anticipated or active periodontal problems in this population.

- **Periodontal evaluation:** Early documentation of periodontal status, especially bone loss and pocket formation, is important. It is quite unusual for most dentists to contemplate documentation of periodontal disease onset and advancement at such an early age (as early 6-16 years old); however, this is an important oral health care consideration in this population. For example, if an extraction is needed in an 18 year-old patient due to advanced bone loss, documentation of the disease progression and therapies attempted will let all involved parties know that the oral health provider was attentive to the patient's periodontal condition.

- **Periodontal scaling and root planing:** Periodontal scaling and root planing should be performed as indicated for patients without Down syndrome.

- **Topical antimicrobial agents:** Topical antimicrobial agents (e.g., chlorhexidine oral solution or Listerine®) may be indicated on a long term basis for some individuals with Down syndrome. The ability of the individual to rinse and

expectorate may be a limiting factor in the use of oral solutions. In those cases, utilizing the solution to moisten the toothbrush bristles prior to brushing may prove to be an effective delivery mechanism. Other delivery methods such as gels or sprays may be helpful for some patients.

- **Systemic and local antibacterial agents:** Systemic and local antibacterial agents, particularly tetracycline may be helpful. Options include low dose doxycycline (Periostat®), penicillin, and metronidazole, among others. A combination of omega-3 DHA and EPA along with aspirin is also being investigated. Local options include a chlorhexidine chip (Periochip®), doxycycline (Atridox®), minocycline (Arestin®), tetracycline fibers (Actisite®), and metronidazole gel (Elyzol®). The major problem is the lack of data confirming the success of periodontal therapy in the presence of an altered immune or host response.

- **Extractions:** Consideration of aggressive treatment, including early extractions of periodontally involved teeth, should be weighed against the risks of early tooth loss. Many individuals with Down syndrome cannot tolerate traditional replacement of teeth with removable or fixed prostheses. However, the need for extractions of periodontally infected teeth, for example teeth with suppurative periodontium which place the patient at risk for systemic infection, is clear. Also, since crowded dentition is associated with increased periodontal problems, selected extraction of primary or permanent teeth may be considered. Since host response is impaired and post-surgical healing may be prolonged, the use of antibiotics is sometimes recommended; however, the evidence is lacking on the need for antibiotics in this population.

- **Grafts:** There is some evidence that some persons with Down syndrome have a peripheral circulation problem with an abnormal capillary or vascular system which could affect the healing and success of autogenous gingival grafts. The use of bone grafts or bone replacement products (e.g., hydroxyapatite) may hold promise for future use.

- **NUG:** Early and aggressive therapy, where there is evidence of NUG or similar dental papilla destruction, is important.

Prosthetic Treatment

Prosthetic treatment choices can be very limited in this population. The existence of periodontal disease and tooth mobility can severely limit the choice of fixed or removable partial dentures. The reduced cooperative ability seen in many patients with Down syndrome at times renders these choices problematical. Complete dentures are often not a viable treatment choice due to lack of retention (for example, poor orofacial neuromuscular control due to oral hypotonia), presence of severe class III jaw relationship and reduced cooperation and/or tolerance by the patient. Relative lack of maxillary sinuses in some of these patients would theoretically aid in the use of implant technology; however, the issues of patient cooperation often preclude this treatment choice. It is imperative that the limitations of prosthetic replacement be fully explained to the patient and guardian early in the treatment planning stage to provide realistic expectations in this regard.

Orthodontics and Cosmetic Surgery

- **Orthodontics:** Reports of orthodontic treatment in patients with Down syndrome show that treatment can be successful in select cases. Early orthodontic intervention with palatal expansion and cross bite correction may reduce the impact of expected

malocclusion. However, many patients with Down syndrome are not able to tolerate routine orthodontic treatment.

- **Cosmetic surgery:** There is considerable controversy regarding the advantages of cosmetic surgery to reduce the stigma of Down syndrome. These procedures would include augmentation of the nasal, cheek and chin areas, glossectomies and lateral canthoplasty. Some parents support the surgery as a normalizing procedure. Opponents contend that undergoing a surgical procedure to correct a societal problem (the stigma of Down syndrome) is too radical and serious a procedure for any possible gains expected. These types of procedures are very rarely done for patients with Down syndrome.

SUMMARY

The provision of dental services for the person with Down syndrome presents unique challenges to dental providers. A thorough knowledge of the dental implications of this syndrome and an innovative problem solving approach to treatment planning and preventive procedures will do much to alleviate the dental effects of this condition.

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