

Case Report**Dental Care of Infants and Young Children with Down Syndrome: A Review**

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ABSTRACT: Down's syndrome is caused by a chromosomal abnormality and is characterized by certain physical, mental and medical features with specific oral manifestations. A number of these features, such as learning disability, cardiac anomalies and an altered immune system and oral manifestations like early onset of severe periodontal disease (most significant oral health problem), lower prevalence of dental caries, delayed eruption of permanent teeth, malocclusion, congenitally missing and malformed teeth, hypoplasia of mid-facial region, hypodontia, microdontia, macroglossia, fissured and protruding tongue, tongue thrust, bruxism, clenching, mouth breathing can have a profound effect on oral health and the delivery of oral care. In turn, this can affect social acceptability and quality of life. The optimum potential of the person with Down's syndrome is achieved via a multidisciplinary approach that involves the pediatric dentist from an early stage.

Keywords: Down syndrome; Trisomy; 21st chromosome; Hypoplasia; Microdontia; Macroglossia.

INTRODUCTION

Down syndrome is one of the most common genetic syndromes, occurring in one of 800 to 1,000 live births.¹ Down syndrome is a chromosomal disorder associated with an extra chromosome (Trisomy 21) resulting in intellectual disability and specific physical features. The teeth of people with Down syndrome, both baby teeth and permanent teeth, may come in late compared to children without Down syndrome. On average, babies with Down syndrome get their first teeth at 12 to 14 months, but it may be as late as 24 months of age. Babies without Down syndrome typically get their first teeth between 6-12 months. It is typical that a child with Down syndrome may not get all 20 baby teeth until he or she is 4 to 5 years of age, rather than 2-3 years of age, which is typical for children without Down syndrome. The front permanent teeth and permanent 6 year old molars may not erupt until 8-9 years of age. It is also common for the teeth of children with Down syndrome to erupt in a different order than in children without Down syndrome.²

Systemic factors influencing dental care

Although 40 to 50% of babies with Down syndrome are born with some type of cardiac

abnormality, most receive surgical correction within the first few years of life. There is however, an abnormally large percentage that develop mitral valve prolapsed (MVP) by adulthood. The incidence of MVP in the normal population is between 5-15%. Approximately 50% of adults with Down syndrome have mitral valve prolapse requiring subacute bacterial endocarditis (SBE) prophylaxis for dental treatment.³ One third of these adults with MVP do not have auscultatory findings, requiring diagnosis of the MVP by echocardiogram. Patients with Down syndrome or their caregivers may not be aware of the need for diagnostic echocardiology in adulthood.

A compromised immune system with a corresponding decrease in number of T cells is characteristic of most individuals with Down syndrome; this contributes to a higher rate of infections and is also a contributing factor in the extremely high incidence of periodontal disease. Children with Down syndrome often have chronic upper respiratory infections (URIs). These contribute to mouth breathing with its associated effects of xerostomia (dry mouth) and fissuring of the tongue and lips. There is also a greater incidence of aphthous ulcers, oral candida infections and ANUG.⁴

A reduced degree of muscle tone (hypotonia) is generally found in Down syndrome. This

affects the musculature of the head and oral cavity as well as the large skeletal muscles. The reduced muscle tone in the lips and cheeks contribute to an imbalance of forces on the teeth with the force of the tongue being a greater influence. This contributes to the open bite often seen in Down syndrome.

Additionally, reduced muscle tone causes less efficient chewing and natural cleansing of the teeth. More food may remain on the teeth after eating due to this inefficient chewing. Associated with the low muscle tone seen in Down syndrome is a ligamentous laxity seen throughout the body. This causes hyperflexibility of the joints and it is theorised that the ligaments around teeth may be influenced as well.⁵ A condition related to ligamentous laxity is that of Atlanto Axial Instability. The diagnosis and significance of this condition is controversial but is described as an increase in mobility between the C1 and C2 cervical vertebrae and may be seen in 10-20% of individuals with Down syndrome. If a patient has this instability, careful positioning in the dental chair is required to avoid any potential harm to the spinal cord.

Persons with Down syndrome vary widely as to their degree of intellectual impairment. Most have IQs in the mild to moderate range and are able to be treated in a normal setting. There is often a relatively severe delay in language development. The patient with Down syndrome will probably understand more than their apparent level of verbal skills. The assistance of the patient's family or caregiver will be necessary in conveying to the dentist and staff what level of communication should be used with the patient. It may take a little extra appointment time to explain procedures to the patient with Down syndrome, but once a level of trust is achieved they are likely to be very co-operative patients.

Down syndrome is frequently seen in conjunction with other medical problems. There is a higher incidence of epilepsy, diabetes, leukemia, hypothyroidism and other

conditions. Alzheimer's disease and Down syndrome appear to have a strong connection to one another. The importance of a thorough medical history including a work-up by a physician cannot be over emphasized

Oral Manifestations:

1. Small and missing teeth: Frequently, people with Down syndrome have smaller than average teeth and missing teeth. It is also common for the teeth of people with Down syndrome to have roots that are shorter than average.

2. Large tongue: People with Down syndrome may have large tongues or they may have an average size tongue and a small upper jaw that makes their tongue too large for their mouth. It is also common for people with Down syndrome to have grooves and fissures on their tongues.⁵

3. Problems with bite: People with Down syndrome may have small teeth, which can cause spacing between the teeth. They also tend to have a small upper jaw. This may cause crowding of the teeth and may result in the permanent teeth being "impacted" because there is no room in the mouth for them to come in. The small upper jaw may create a situation where the top teeth do not go over the bottom teeth the way they are meant to; instead, the bottom teeth may be out further than the top teeth in the back of the jaw, the front of the jaw, or both. It is also common that the front teeth of people with Down syndrome do not touch. Orthodontics (braces) may be able to improve some of these issues. Orthodontics require a lot of cooperation and make the teeth even more difficult to keep clean, so it may not be possible in all people. It may be a good idea to wait until a child is older and able to tolerate it a bit better. Having orthodontic appliances in the mouth can also pose challenges to speech. Children without Down syndrome typically adapt their speech quickly; however, in a child with Down syndrome, where speech may already be an issue, adapting to the appliances may be very difficult. Therefore, it may be a good idea to delay orthodontic treatment until a

child is older and his or her speech is further along.

4. **Gum disease:** People with Down syndrome are at an increased risk for gum disease (periodontal disease). Even when individuals with Down syndrome do not have a lot of plaque and tartar (calculus), they get periodontal disease more frequently than others. This is because people with Down syndrome have an impaired immune system and do not have some of the natural protections against the disease that people without Down syndrome have. To prevent gum disease brush twice daily, focusing the bristles along the gum line, floss daily and be sure to visit the dentist regularly to have gum health monitored and to take X-rays to monitor bone levels. If the gums bleed that means that they are inflamed. Brushing and flossing should not be stopped because of this. In fact, brushing and flossing will keep the gums clean and help to minimize the inflammation.

5. **Cavities:** Some research says that people with Down syndrome are at less of a risk for cavities; however, much of that research was done when people with Down syndrome lived in institutions and had very restricted diets. People with Down syndrome do get cavities, so brushing with fluoride toothpaste, flossing between any teeth that touch, and limiting the amount and frequency of sugar and refined carbohydrates eaten will help to prevent the development of cavities.⁶

Treatment Objectives

Treatment objectives for any population with developmental disabilities should be the same as that of normal patients. Treatment plans may need to be adapted as necessary due to each individual's condition, but the overall goal should be to provide as comprehensive treatment as possible. Areas of dental care such as Pedodontics, cosmetic dentistry, orthodontics, prosthodontics, and reconstructive oral surgery should not be ruled out simply because the patient has Down syndrome. With the numbers of persons with Down syndrome working and living out in the community, there may be many who desire

and can handle some of the more extensive dental treatment options available today.

Behaviour Management

Good behavior in the dental office is learned. In a population with delayed learning, this can be a challenge for the dentist and staff. Dental treatment for children with Down syndrome may not be sought out at an early age. There may be more pressing medical problems, financial considerations or parents may want to wait until the child seems mature enough to handle a visit to the dentist. Unfortunately this makes it more difficult to teach proper home care and to develop a relationship with the child that will result in co-operative behaviour during dental treatment.⁷

Many children with Down syndrome can successfully be treated in the dental office.

Guidance

- Plan a pre-appointment (in person/ phone) to discuss patient special needs prior to the first visit. Discuss this with the parent or care provider-they know the child best.
- Schedule appointments early in the morning or best time of day for patient.
- Talk with the parent or caregiver to determine the patient's level of intellectual and functional abilities and explain each procedure at a level the patient can understand.
- Use short, clear instructions and speak directly to the patient.
- Minimize distractions, such as sights and sounds, which may make it difficult for the patient to cooperate.
- Start the oral examination slowly, using only fingers at first. If this is successful, begin using dental instruments.
- Use the Tell-Show-Do approach when introducing new instruments or procedures.
- Reward cooperative behavior with positive verbal reinforcement.
- Develop trust and consistency between the dental staff and the patient. Use the same staff, dental operatory, and

appointment time each visit if appropriate.⁸

Dental Treatment And Prevention:

- Consider patient's cardiac status and need for premedication-medical consult may be indicated
- It is not uncommon to encounter patients who are tube-fed among the population of Children with Special Healthcare Needs. Patients fed by tube typically have low caries, rapid accumulation of calculus, GERD (Gastro-esophageal Reflux Disease), oral hypersensitivity, and are at high risk for aspiration in the dental chair. No antibiotic premedication is needed for Gastric or Nasogastric tubes. Position the patient in as upright a position as possible and utilize low amounts of water and high volume suction to minimize aspiration.
- Examine patients by the first birthday; monitor tooth eruption patterns and malformations.
- Monitor periodontal disease. Treat as needed and consider specialty referral if indicated.
- Powered toothbrushes may be too stimulating for some children and should be recommended only after determining if the child will tolerate one.
- Consider prescribing Chlorhexidine or other antimicrobial agents for daily use.
- Some patients are good candidates for full orthodontic treatment. Maintain primary teeth as long as possible and consider space maintenance and orthodontic consultation for missing teeth.⁹

Considerations due to Associated Medical Conditions:

- Increased gag reflex during oral examination.
- Chronic respiratory infections and open mouth posture frequent mouth breathing xerostomia.

- Seizure management during treatment: Remove all dental instruments from the mouth. Clear the area around the dental chair. Stay with the child and turn child to one side. Monitor airway to reduce risk of aspiration. Note time seizure begins: if seizure continues >3 min call EMS – Danger of Status Epilepticus (potentially life threatening).
- Carefully move patients with atlantoaxial instability into the dental chair, giving special attention to the spine and neck. Use pillows to stabilize the patient and increase comfort, as directed by the caregiver.¹⁰

CONCLUSION: Life tables published in 1989 showed that more than 50 percent of infants with Down syndrome could be expected to live more than 50 years.¹¹ As mortality rates for the operative repair of congenital heart defects continue to decrease, survival may increase considerably.¹² Since the trend toward deinstitutionalization began in the 1970s, it has become apparent that the maintenance of optimal health is a major factor in the lifelong functioning of persons with Down syndrome. Primary health care has become essential to helping these persons have longer, more productive lives. Dental care for the patient with Down syndrome can be achieved in the general practitioner's office in most instances with minor adaptations. Although this population has some unique dental care needs, few patients require special facilities in order to receive dental treatment. Adequate dental health care for persons with developmental disabilities is a major unmet health need. It is hoped that the information contained in this review will encourage general practitioners to be willing to provide comprehensive dental care to their patients with Down syndrome.

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