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Early Dental Intervention in Children with Down Syndrome – A Review

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Abstract

Down's syndrome is a condition which is associated with a chromosomal abnormality. It is usually in the form of three in place of a pair of chromosomes 21, hence the name Trisomy 21. These children have mental retardation to a certain degree and they are generally smaller than average in height for their age. Flat broad bridge-shaped nose, midface dysplasia, "lop" ears, epicanthal folds with slanting almond-shaped eyes are present in such children which are responsible for the term 'Mongoloid'. Early dental intervention should be considered important in such children. This article focuses on the importance of early dental intervention and strategies to combat the same.

Keywords: Down syndrome, Early dental intervention, Dental caries, Periodontal condition, Oral hygiene

Introduction

Down syndrome was first described by John Langdon Down in 1866. He recognized the phenotypic expression of patients with circulation and coordination problems as having down syndrome. A century later, Jerome Lejeune hypothesized that non-disjunction during meiosis could lead to Trisomy of the 21st chromosome. The incidence of Trisomy 21 (T21) is 1 in 800 to 1000 live births in the US.[1,2] There are three types of Down's syndrome[3] –

i) Trisomy 21 (94%) – The extra 21 chromosome (3 instead of 2) produces a complement of 47 chromosomes.
ii) Translocation (5%) – A segment of a 21 chromosome is attached to other pair of chromosome (usually #14, thus referred to as 14/21 translocation). A normal complement of 46 chromosomes is present in such individuals.
iii) Mosaicism (1%) – Non-disjunction may occur at a later stage of cell division. Some cells have the normal complement of 46 chromosomes and other cells have 47 chromosomes (with an extra 21 chromosome).

Epidemiology

Down syndrome is usually in the form of three in place of a pair of chromosome 21, hence the name "Trisomy 21". This condition is more frequent in children born to mothers who are aged 35 years or above. In young mothers, there may be an inherited translocation of one of the pair of chromosome 21 which may become a third in the offspring, resulting in Down syndrome. The incidence of this syndrome in the population has been variably estimated between 1 and 4 per 1000.[4]

Facial Features[5,6]

Moderate to severe mental retardation is seen in all Down's syndrome patients. They have

delayed milestones of development. They have a flat broad nose with deviated nasal septum and narrow air passage. The ears are low-set with a flat or absent helix. They have slanting almond-shaped eyes with narrow palpebral tissue slanting towards the midline which was responsible for the term 'mongoloid'. An epicanthus is often present. Other eye abnormalities are strabismus, in more than half of the patients, nystagmus and lens opacities. They have a broad and short head (brachycephaly). There is absence of supraorbital ridges. There is hypotelerism secondary to hypoplasia of the central face. Frontal sinuses are absent. There may be absent or reduced maxillary sinuses. The mouth is open with a protruding tongue. The neck is broad and short. The chest has flat nipples. The abdominal abnormalities are diastasis recti and umbilical hernia. The hands are short and broad with single palmar crease. The fifth finger is short. They have hyper flexible joints.

Medical Conditions

The patients with Down's syndrome have a wide array of signs and symptoms like intellectual and developmental disabilities or neurological defects, congenital heart defects, gastrointestinal abnormalities, hematological, endocrinological, musculoskeletal and visual disorders. There may also be disorders of thyroid gland and hypopituitarism. Down syndrome patients are prone to infections that result in bronchitis and other respiratory infections. There is also higher incidence of leukemia in such patients.[7]

Oral Manifestations[7,8,9]

About one-third of these patients may have congenitally missing teeth. The wrinkled occlusal surfaces of molars are believed to occur during tooth development. Other oral manifestations are listed in Table 1.

Oral Conditions

There are mainly three widely common oral conditions seen in such patients. Dental caries: Children with Down syndrome have remarkable resistance to dental caries.[10] This may be due to the simpler form of teeth with few deep fissures. The high frequency of hypodontia and delayed eruption patterns have been proposed for the low prevalence of dental caries.[11] According to analysis of electrolytes in oral secretions, increased pH concentrations of sodium chloride and bicarbonates contribute to an increased buffering effect thereby reducing the risk of dental caries.

Periodontal condition:

Periodontal disease is more severe in patients with Down syndrome. It is more frequent in the mandibular incisor area. Even at the age of 3 years there may be tissue breakdown leading to early loss of primary central incisors. Due to limited access to care, limited manual dexterity and lowered efficacy of self home care, there is increased levels of gingivitis.[12] Radiographically, there is unclear lamina dura and the bone trabeculae appear shorter and thicker with consequently smaller narrow spaces. Most of the patients have tongue thrusting habit with lack of lip seal, high frenal attachment or in tight contact and traumatic occlusion could be contributing factors for periodontal disease.[13]

Malocclusion:

The presence of underdeveloped and retrognathic maxilla and a relatively large and prognathic mandible contribute to the development of Angle's Class III malocclusion.[14] This occurs in one-third or more of these children. The occurrence of malocclusion is listed in Table 2.

Table 1:

No of Teeth	Congenitally missing teeth Hypodontia One or both missing maxillary lateral incisors (more frequent)
Morphology	Small, peg-shaped laterals Enamel hypoplasia Rounded, bulbous, short roots Varied fissure patterns Wrinkled occlusal surfaces of molars
Eruption	Delayed
Tongue	Protruded

Table 2:

Malocclusion	**Occurrence**
Posterior cross-bite	Bilateral
Anterior cross-bite	Unilateral
Anterior open-bite	62% in boys 48% in girls
Posterior open-bite	More common in females
Reversed anterior overjet	Proclined mandibular incisors
Anterior crowding	30%
Posterior crowding	39%

The posterior and anterior crossbites may be due to increased widths of mandibular arch as compared to maxillary arch. The etiological factors that are associated with open bite is deficient maxillary growth accompanied by tongue thrust. There is often lack of lip seal that result in proclination of the mandibular incisors, accentuating the reversed incisor relationship.[15] The crowding of teeth may be attributed to the tongue function, posture and size of the maxillary antero-posterior and vertical growth insufficiency.

Due to the delayed emergence of the permanent teeth and the high number of missing teeth, it is crucial to maintain the primary dentition as long as possible.[16]

Early Dental Intervention

The American Dental Association (ADA) recommends to make an appointment with the dentist after the first tooth erupts in the oral cavity but not later than 1 year of age.[17] Early dental intervention is crucial in recognizing potential problems and more importantly teaching children how to care for their teeth and gums for lifetime. Starting early is the best way to give children with Down syndrome a roadmap towards overall dental health.

Importance Of Early Dental Intervention

- To prevent dental caries and periodontal disease
- To promote good oral hygiene habits
- To manage and prevent malocclusion complications
- To encourage parental involvement in oral care
- To provide a positive dental experience to reduce anxiety

Early and Preventive Dental Treatment Considerations

Oral health is an integral part of total health, not an isolated element. Prevention of oral disease is the key to the oral care of Down syndrome patients. Because children with

Down syndrome have a higher risk of dental health problems, prevention of the same would be beneficial for the patient.[18]

Pre-Treatment Assessment –

The dentist should take proper medical history and should have proper consultation with their physician to evaluate their medical status. Information about the patient's oral hygiene practices should be collected from their caretakers or guardians at the time of first appointment only.[19]

Patient Management –

An appropriate method of behavior management must be determined. Children with Down syndrome are usually cooperative. Compliant issues should be dealt with the same way as far any other children presenting for dental treatment.[20] A relaxed environment must be established. Communication should be done in a soft voice and using a gentle touch. Tell-show-do behavior modification technique is usually implied in such patients. An adjustable dental bite; a mouth prop may be used.

Preventive Protocol –

Oral health education should have Specific, Measurable, Appropriate, Realistic and TimeRelated (SMART) objectives. These objectives must include policy development, improved availability of healthy choices, improvements in oral hygiene skill and provision of services.[21,22]

1. **Plaque Control** – It can be done by mechanical means or chemical means.

Mechanical means: Toothbrush is an effective way to remove plaque. Patients who are not able to handle it properly need help of a caregiver. Powered toothbrushes are designed to clean the oral, buccal and occlusal surfaces of the teeth with a single stroke and is recommended for certain individuals with limited manual skills.[23]

Chemical means - Chlorhexidine is the treatment of choice for gingivitis. It acts as an adjunct for patients who are unable to remove plaque by mechanical means.[24] For patients who are unable to use chlorhexidine as mouthwash, the agent can be swabbed on the teeth with an applicator, sprayed on the teeth, applied with a toothbrush or used as a gel.[25]

2. **Pit and Fissure Sealants** – These are thin, protective coating applied to the occlusal surface of premolars and molars. They act as physical barrier, thus protecting the tooth enamel from plaque accumulation and acid attacks that can lead to development of dental caries. A study by William et al. found that after 1 year, 74% of sealants were fully retained without any caries.[26] A 14-year old follow-up study of Balian found that dental sealants were 93.6% effective in preventing tooth decay.[27] Thus regular monitoring and maintenance of pit and fissure sealants is required.
3. **Fluoride –** The benefits of fluoride in the prevention of dental caries is well documented. Optimized fluoride in drinking water remains the cornerstone of prevention.[28] For children aged 6-16 years, 1mg/day of fluoride supplement should be used if fluoride concentration in drinking water is under 0.3 ppm. 0.5 mg/day of fluoride supplement should be given if the concentration of fluoride in drinking water is 0.3-0.5 ppm. If the fluoride concentration in water exceeds 0.5 ppm, no fluoride supplement is needed. An appropriate fluoride concentration in toothpaste is very crucial factor in preventing the risk of dental caries. 1000-1500 ppm of fluoride is a recommended level in toothpastes. In patients who are unable to rinse off the paste from teeth, parents should brush their teeth and wipe off the foam.[29]

For professional use, fluoride varnish is the most ideal preventive dental agent for children. Toothbrushing should be done twice daily with

a soft toothbrush with fluoridated toothpaste. Children under 3 years of age, a thin 'smear' of toothpaste should be used whereas, for children above 5 years of age, 'pea-sized' amount of toothpaste is advisable.

4. **Scaling and Prophylaxis –** Stains and calculus should be removed by ultrasonic scaling by a dentist. For children with congenital heart disease, deep scaling should be done under antibiotic coverage.[30] The periodontal condition constitutes the principle difficulty in attaining any degree of one's dental health. Therefore, periodic regular dental checkups, i.e.; once in 3 months would be beneficial for the patient.
5. **Orthodontic and Prosthodontic Treatment –** Both orthodontic and prosthodontic appliances are not commonly used in children with Down syndrome due to poor gingival condition, relatively large tongue and poor muscle tone. The short roots are disadvantageous for orthodontic tooth movement. Removable prosthesis may be considered a contraindication due to inadequate cooperation.[31]
6. **Diet and Eating Behavior –** Children who consume sugary, starchy and sticky foods are at a high risk of developing dental problems. Streptococcus mutans and Lactobacillus species which are present in dental plaque sugar producing organic acids. The decreased pH on the tooth surface leads to demineralization and eventually cause dental caries.[32] Night time bottle-feeding leads to tooth decay due to prolonged presence of milk sugar on the teeth.[33] Therefore, food and beverages with added sugars should be avoided. A well balanced-diet rich in fruits, vegetables, whole grains and lean proteins is crucial for maintaining oral health, thus maintaining general health.
7. **Bruxism –** Custom-fitted mouth guards should be provided for children who are at risk of tooth damage from bruxism or trauma.[34]

School Oral Health Services

- Educational institutions should include oral health education as a part of training programs.
- A dental health council should be created that include teachers, parents, community leaders, dental professionals, etc. as a part of school community
- In service-training to promote good oral hygiene for children with disabilities.
- Specific programs should be established such as toothbrushing education campaigns, classroom-based fluoride rinsing programs, diet counseling, etc.[35]

Behavior Management[36]

- Most children with Mongolism can be successfully treated in the dental office.
- Schedule appointments in the morning.
- Talk with the parent/caregiver to determine the child's level of intellectual abilities and explain each procedure at a level the child can understand.
- Use of short and clear instructions.
- Minimize distractions, such as sight and sounds that may make it difficult for the patient to cooperate.
- Use of Tell-Show-Do approach when introducing new instruments or procedures
- Reward cooperative behavior with positive reinforcements.
- Use of physical restraints such as papoose board.

Conclusion

Early dental intervention and proper preventive protocols should be given higher priority in children with Down syndrome. Oral health promotion should include facilitating access and regular use of oral health services. Early dental intervention and its proper implementation not only prevents oral diseases

but also teaches the child to maintain good oral hygiene.

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