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RESEARCH ARTICLE

PREVALENCE OF MALOCCLUSION IN DOWN SYNDROME POPULATION

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ABSTRACT

Down syndrome (DS) is a genetic disorder associated with various craniofacial abnormalities, including malocclusion. Understanding the prevalence and types of malocclusion in individuals with DS is crucial for developing tailored orthodontic treatment plans and improving their oral health and quality of life. This study contributes valuable insights into the orthodontic management of DS patients and emphasizes the importance of multidisciplinary care in addressing their unique dental needs.

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INTRODUCTION

DOWN SYNDROME also known as TRISOMY 21 is a congenital disease arising due to defect in chromosome, which results in intellectual impairment and physical abnormalities. Down syndrome is the most common cause of intellectual disability among livebornchildren. It was first described by Langdon John Down, a British physician who practiced at London and Earlwood, according to him, it is a unique physical and mental characteristics of a subgroup of intellectually challenged individuals under his care. it is a gene defect with an additional chromosome in the Chromosome 21 at birth or before birth and is a lifelong condition. Different clinical conditions are associated with Down Syndrome as different systems are affected by it. The present review is aimed to evaluate the prevalence of dental malocclusion in Down syndrome individuals. Various screening tests are recommended to married couple to determine the risk of Down syndrome fetus. The extent of dental and skeletal malformation can be identified using various indices, Angle's classification and cephalometric analysis and high rate of findings requiring treatment in patients with Down's syndrome represent a significant indication for orthodontic therapy.

Down Syndrome: Down Syndrome is a rare genetic condition that occurs when there's an unnatural celldivision due to replication of chromosome 21. Although the conditions may manifest in various degrees, there are certain common symptoms that all patients, regardless of age will display. The peculiar aspect of these subjects is partly as a result of developmental anomalies of craniofacial skeleton.

Subsequently the group of individuals were referred to as Mongoloid and often the disease was called Mongolism. But in 1965, World Health Organization dropped the term Mongolism and in 1975 National Institute of Health, USA, recommended the term DOWN SYNDROME to replace the other names that described such a phenotype. Chromosomal disorder occurs in 1 out of 166 live birth in India out of which 1 out of every 830 children being born with Down Syndrome. Out of 23, 000 to 29,000 kids born with Down Syndrome in India every year the survival rate is only 44%. Maternal age is strongly associated with the prevalence of chromosomal anomalies especially in Down syndrome. Shuttleworth (1909) was the first to identify the relative advanced maternal age of the mother at the time of gestation.[1] Followed by Penrose in 1933 established the importance of advanced maternal age in the genesis of Down syndrome [2]. In 1975, Verma and Singh examined over 600 cases of Down syndrome in various parts of India and found that women over 35 years formed 33% of the mother's giving birth to children with Down Syndrome.[3]Kochupillai et al in 1976 have demonstrated only that mother's in the age group 30-39 year run a risk 10 times greater than those in younger age group. According to recent studies, the risk increases with mother's age (1 in 1250 for a 25 year old mother to 1 in 1000 at age 31, 1 in 400 at age 35, and about 1 in 100 at age 40). Clinically, it is characterized by neurological changes, structural cardiopathy, respiratory problems, increased risk of infection, increased risk of Leukemia, dental anomalies, craniofacial disturbances, bone growth disorders and generalised muscle hypotonia. The trisomy 21 is marked by craniofacial characteristics that make it well defined and readily identified. Prenatal testing is the best strategy for reducing the burden of genetic disorders and

congenital disabilities that can cause significant postnatal functional impairment. With advanced medical knowledge, screening tests for genetic disorders are available for all women before marriage or before conception. Such screening before marriage and before conception can help couple to know whether their child will be at use of any inherited or genetic ailment or not. These test carry no risk of miscarriage and detect the likelihood of a baby being born with disorders[4,5] Early detection through screening allows parents to be better prepared for the possibility of having a child with special needs.This information is vital for making informed decisions about the pregnancy, including potential medical needs and care planning. Knowing in advance enables medical preparations and the arrangement of specialized care and interventions.Counseling and support help parents emotionally and psychologically prepare for the challenges and joys of raising a child with Down syndrome. Counseling provides emotional support, which is essential for coping with the potential diagnosis and the emotions it may evoke.Information provided through counseling empowers parents with knowledge about Down syndrome and available resources, aiding them in making informed decisions. Counselors play a crucial role in connecting parents to support groups, early intervention programs, and experienced medical professionals, ensuring the child receives the best care andopportunities.Additionally, counseling can strengthen the parental bond and prepare them for the unique experiences that come with raising a child with Down syndrome.[6]

Screening tests

Carrier Screening: Genetic testing that is performed on an individual who doesn't have any overt phenotype for a genetic disorder but can have an allele within a gene that is associated with the syndrome /disease. This enables couple to understand their reproductive risk.

Mutation screening: Mostly done in case of genetic condition that has already been diagnosed or has a family history. Usually done in conjunction with clinical presentation.

Non Invasive Prenatal testing: This test is extremely sensitive to chromosome 13,18,21.

Amniocentesis: For this test, amniotic fluid is collected from the uterus as this fluid contains fetal cells and various proteins.

Chromosomal Microarray analysis*:* This analysis detects clinically significant microdeletion or duplications.

Various other tests include Karyotyping, Double Marker test, Triple Marker test, Quadruple test, FISH, QF-PCR.

Oral manifestations

The orocraniofacial features includes brachycephaly,low nasal bridge, small maxilla tongue with fissures, papillary hypertrophy prominent epicanthic folds,hypoplastic cranial base, hypoplastic facial middle third, smaller lower facial middle third. Some of the dental features includes taurodontism, openbite, macroglossia, hypodontia, microstomia, anondontia, drooling, poor oral hygiene, high caries risk, delayed eruptions, small maxilla retained deciduous teeth, bruxism. The high prevalence of dented anomalies is associated with slow cellular growth rhythm and reduction in number of cells that shows strong influence on tooth development. The tooth eruption can be delayed or may occur in an unusual order and this causes the tooth to erupt in gingival tissues at atypical sites causing malocclusion.[6]

Occlusal Anomalies

Normal alignment of teeth not only contributes to oral heaith but also goes a long way in the overall will being and personality of the individual. Correct tooth positioning is an important factor for function, esthetics and for overall preservation of dental health. Many organized population survey were carried out in different parts of the world to analyse the prevalence of malocclusion and the treatment needs. The prevalence of malocclusion among Indian Population has been reported to be between 19.6% and 90%, in Delhi with high predominance of Class I malocclusions in studies conducted in various parts of india. [8,9] A higher prevalence of dental malocclusion were suggested among individuals with Down Syndrome compared to non Syndromic individuals. Class III malocclusion was more prevalent among Down's syndrome Population followed by class II malocclusion and class I malocclusion. Early intervention for the correction of malocclusion using face mask and palatal expansion therapy for class III and functional appliances for class II is suggested. [10] There is evidence indicating the premature closure of the suture in the labiaI region resulting in the maxillary deficiency that leads to class III malocclusion.[11] There are significant difference in cephalometric findings between individuals with Down Syndrome and without Down Syndrome.Individuals with Down Syndrome they tend to have a shorter anterior cranial base , maxillary length, upper anterior facial height and mandibular height also the ANB and SNB angles were also smaller while SNA angle showed no difference , which indicated that class Ill tendency was not due to mandibular prognathism.Gonial angles, mandibular plane angles and airway measurements increased with age.[12,13] According to Cohen and Winter, their study revealed increased percentage of anterior open bite following the increase in tongue thursting habit. The prevalence of open bite and crossbite was also associated with the use of bottle feeding and pacifier sucking [14]. The relevance of using IOTN-DC index among DS population way to assess the and of orthodontic treatment by determining the malocclusion features, it had shown severe-to-very severe malocclusion, in different researches, indicative of mandatory need for orthodontic treatment[15].

Treatment Considerations

There should be a high orthodontic treatment need for DS patient because of increased prevalence and severity of malocclusion, nevertheless the treatment seevices have been generally neglected, even though they are willing to undergo treatment. The treatment procedures need to be simplified so it is possible to be done; the treatment objective have to became as a normal patient while the treatment plan have to be adapted to each individuals condition.

Palatal plate: Muscle hypotonia, a characteristic of individuals with Down syndrome, impairs orofacial development , causing functional limitation in sucking, breathing , mastication and speech.The use of an intraoral appliance named stimulating palatal plate (SPP) in combination with Orofacial Regulation Therapy was proposed by Castillo - Morales in 1970s. This study conducted was conducted in children between 6 months of age to 48 months of age showed that treatment had a positive effect on oral motor function, highlighting improved tonus and lip closure[16].

Palatal expansion: Maxillary Expansion can be undertaken in Down Syndrome children, and is succesful in high proportion of cases if correct case selection has been done. Slow activation during expansion phase is preferred since they are more susceptible to appearance of oral ulceration which can affect the negatively on the course of treatment.[17]

Fixed Orthodontic Treatment: Little literature is available on orthodontic treatment in patients with down syndrome, particularly regarding the use of fixed appliances. Abeleira et al. investigated in a case-control study the use of fixed multibracket dental therapy in which it was reported that, in patients with down syndrome, orthodontic treatment takes longer than usual also the frequency of complications is higher than in the general population.[18,19] In case of fixed orthodontic treatment,bracket placement should be performed indirectly for increased accuracy and reduced treatment time. The most time-consuming steps, including those in the dental laboratory, should be completed outside the patient's mouth. Light forces should

be applied to move the teeth to avoid patient discomfort. The straight wire technique is a suitable choice due to minimal wire changes. Self-ligating brackets are beneficial for reduced appointment times and improved oral hygiene. Fixed appliances should only be worn for a limited period, and proper bonding is essential to prevent debonding. Replacement of brackets is time-consuming. Procedures like wire changes and activations can be uncomfortable due to the numerous instruments used in the mouth. In non-cooperative Down syndrome patients, fixed appliances are necessary since they cannot remove them on their own. However, their immature condition may lead to appliance damage and oral mucosal injuries. After active treatment, a retention period is essential to maintain the results. Skeletal discrepancies or large tongues, non-eliminated bad habits, and non-compliant patients can lead to relapse. Removable retainers like Hawley, wraparound, or clear retainers may be used, depending on patient cooperation. Permanent retainers like bonded lingual retainers are an option for doubtful cooperation.[20]

Mini implants: There are evidences suggesting the use of mini-implants following Rapid Maxillary expansion for the correction of class II dental relationship with a sliding jig[21].

Management in Dental Clinic

While individuals with Down syndrome typically exhibit a cooperative demeanor, ensuring adequate oral hygiene hinges on the family's knowledge and education. Children with Down syndrome may harbor anxiety or fear of dental visits, often unknown to parents. The effectiveness of dental treatment relies heavily on the dental professional's behavior management skills, considering the child's mental, emotional, and social development. Utilizing the tell-show-do technique is usually effective with affectionate and cooperative Down syndrome children. Factors such as prophylaxis for subacute bacterial endocarditis and patient compliance should be taken into account during treatment. Gag reflex can be mitigated through behavioral techniques, intraoral massage, or pharmacological/non-pharmacological interventions. Regular recalls and preventive dental measures are integral to the treatment plan, emphasizing caregiver education for optimal oral hygiene. Mild sedation may address moderate anxiety, while highly resistant patients may necessitate general anesthesia. Give the family a briet tour or the place berore attempting treatment.Introduce the patient and family to the office staff. This will familiarize the patient with the personnel and reduce the patient's fear of the unknown.Allow the patient to bring a favorite item (stuffed animal, blanket or toy to hold for the visit). If the child has an alternative communication system, such as a picture board or electronic device, be sure it is avallable to assist with dental explanations and instructions.

Actively listen as speaking might be challenging for individuals with Down syndrome, demonstrating understanding. Assess the patient's intellectual abilities through communication with caregivers and explain procedures at a suitable level. Provide extra time for oral health discussions and use simple, repetitive instructions to address short-term memory challenges. Collaborate with caregivers to understand effective behavior management techniques, sharing and adopting strategies that motivate the patient. Schedule appointments early in the day for heightened attentiveness and reduced waiting time. Involve the entire dental team to create a positive experience, and minimize distractions during oral care. Conduct a step-by-step evaluation, starting with seating, progressing to an oral examination with fingers, and then incorporating dental instruments. Be consistent in all aspects of care to enhance familiarity and cooperation. Encourage and compliment cooperative behavior, reserving immobilization techniques for necessary situations. Discuss preventive measures with patients and caregivers, emphasizing regular dental appointments and daily oral care. Highlight the importance of fluoride to prevent dental issues associated with xerostomia. Ensure a clear path for movement during treatment, considering the patient's position and spinal cord protection. Maintain visual contact, eliminate background noise, and adjust speaking volume for individuals with hearing impairments. Determine the level of assistance required for safe movement through the dental office. Utilize other senses, like tactile feedback, to establish trust and enhance the overall treatment experience. Keep patients informed about each step, particularly when water is Involved, using clear, descriptive language. [22,23]

CONCLUSION

The stereotype that people with Down syndrome are limited in intellectual and physical abilities is not accurate. In reality, their capabilities vary widely, and many can lead fulfilling and independent lives.Contrary to the misconception that having a child with Down syndrome is an overwhelming burden for families, the experience often involves deep love, joy, and challenges.Many individuals with Down syndrome have the capacity to acquire new skills, attend school, and participate in various activities. While some may require support, many can live independently or with varying levels of assistance.Focusing solely on the health challenges associated with Down syndrome is insufficient. It is important to acknowledge their strengths, interests, and contributions as well.Orthodontic treatment for children with special needs, especially those with systemic disorders and mental challenges, requires simplified procedures to ensure feasibility. Parents often have a strong motivation to seek orthodontic treatment for their children with special needs, aiming for improved appearance and oral hygiene to enhance their quality of life.Patient motivation for orthodontic treatment may not always be clear, but it's essential to recognize that treatment outcomes can be influenced by patient limitations.Down syndrome is not an absolute contraindication to orthodontic treatment, but careful consideration and special care are necessary, especially in patients with systemic, mental, and behavioral conditions.The mental condition of patients significantly impacts orthodontic treatment procedures, and parental support can be beneficial for their psychological well-being.Effective communication with patients with Down syndrome, who may have below-average intelligence, is crucial for establishing a good doctor-patient relationship. Explaining each step clearly and using a "tell-show-do" technique is important to alleviate their apprehension. Behavior disorders can be challenging in treating Down syndrome patients due to limited understanding, increased anxiety, short attention spans, and limited tolerance. Operators should proceed slowly and prioritize patient comfort.Orthodontic treatment is possible if the patient is cooperative, as it involves multiple visits and extended periods. Keeping appointments, maintaining oral hygiene, and avoiding certain foods are important for successful treatment.Parents and caregivers play a crucial role in ensuring good oral hygiene, managing dietary habits, and attending routine dental appointments. Unforeseen circumstances, such as illness, uncontrolled behavior, and inadequate oral hygiene, can lead to treatment failures. In such cases, it is advisable to remove the appliance to prevent further negative effects, even if the desired treatment outcome has not been achieved. It is evident that while Down Syndrome presents challenges, it should not be viewed as surmountable obstacle with right approach and specialised care, individuals with Down syndrome can lead fulfilling lives and benefit from both appropriate medical and dental support.Successful orthodontic treatment not only improves aesthetics but also enhances functions like swallowing, speech, and mastication. Failures can occur due to unpredictable conditions such as illness, uncontrolled behavior, and inadequate oral hygiene, warranting the removal of appliances. Orthodontic treatment remains a possibility for selective Down syndrome patients, though challenges may arise. It requires considerations of medical, mental, and behavioral conditions in addition to the malocclusion itself. Skilled orthodontists, appropriate facilities, and specific efforts, including sedation equipment and trained operators, are essential to achieve treatment objectives.

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