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CASE REPORT

DENTAL MANAGEMENT AND ORODENTAL FEATURES OF A CHILD WITH DOWN'S SYNDROME

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ABSTRACT

Down syndrome (DS) was first described by John Langdon Down (1866) at mid-nineteenth century and one century later, the DS primary cause due to trisomy 21 was reported (Lejeune *et al.,* 1959). DS is predominantly due to non-disjunction of chromosome 21. It is not a disease; however the affected individuals have a greater risk in acquiring many systemic and oral conditions. DS is the most common autosomal chromosomal anomaly with an incidence of 1 in 600 to 1000 live births in all races and economic groups. The extra genetic material in these patients causes a delay in the way a child develops, either mentally or physically. The present article discusses a case report of a 5-year old male patient with the classical features of Down syndrome. The skeletal and soft tissue features, aberrations in dental, periodontal and caries characteristics are discussed and dental management of such patients.

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INTRODUCTION

Down syndrome (DS or DNS) or Down's syndrome, also known as trisomy 21, is a genetic disorder caused by the presence of all or part of a third copy of chromosome 21. Down syndrome is named after John Langdon Down, the British doctor who first described the condition in 1887 [1,2].The incidence is reported to be 1 in 600 to 1 in 700 live births; however, more than half of the affected fetuses spontaneously abort during early pregnancy. Like most trisomies, the incidence of trisomy 21 is highly correlated with maternal age, increasing from about 1/1500 live births for women 20 years of age to 1/30 for women 45 years of age and older. The features of Down syndrome can range from mild to severe. Usually, mental development and physical development are slower in people with Down syndrome. Down syndrome patients have IQs that fall in the mild to moderate range of mental retardation. They may have delayed language development and slow motor development and a characteristic set of facial features (Regezi, 1989; Muller and Young, 2001; Vogel and Motulsky, 1996). This syndrome can be identified in a newborn by direct observation or in fetus by prenatal scanning.

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CASE REPORT

A 5-year-old male patient, first birth order, born to a consanguineous couple aged 33 and 37 years, reported to the Department of Oral Medicine & Radiology with the chief complaint of decayed teeth in the upper as well as the lower teeth regions. The child was diagnosed with Down syndrome by his physician and was not on any medication. His personal history revealed that he brushes once daily with a toothbrush & toothpaste in a circular motion supervised by his mother. Patient had delayed milestones of growth and immunization schedule was completed. Patient attended school with normal kids and was not able to read or write well at school as revealed by his parents. On general examination, patient was moderately built and nourished; his skin appeared to be dry and rough and had mental retardation. Patient is a mouth breather. Patient had most of the common dysmorphic features of Down syndrome. Extra oral examination demonstrated a prominent forehead, brachycephalic skull, saddle nose deformity, retruded maxilla and protruded mandible. Lips were incompetent, appeared rough and cracked. Eyes were almond shaped, ocular hypotelorism, and strabismus was present. Patient had an open mouth posture with slurred speech. Intraoral examination revealed macroglossia with fissures seen on the dorsum of the tongue, high arch and V shaped palate, generalized spacing of teeth in relation to the maxillary and mandibular arches, clinically missing 52, root stumps irt 54, 61, dental caries irt 53 55, deep dental caries with tenderness

associated with 54, 85, teeth were hypodontic and microdontia was present, 73 was conical in shape. Occlusion showed mesial step terminal plane with anterior open bite.

Fig. 1a.

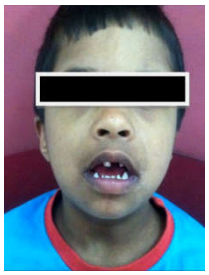


Fig. 1b.

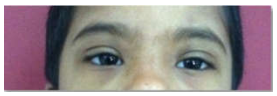


Fig. 1c.

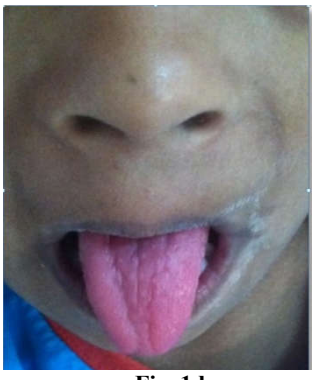


Fig. 1d.

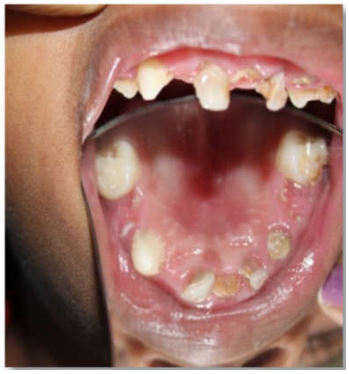


Fig. 1e.

Radiographic diagnosis was not done and was scheduled for the following visits, as the patient was non cooperative hence was graded as having a Negative behavior on Frankel behavior rating scale. The patient was referred to Pedodontics department for the further dental treatment where, the following planning was scheduled:

As the patient was considered to be having a negative behavior, firstly non-invasive treatment was done which were to be followed by invasive treatment, only after obtaining a physician's consent.

Non- invasive procedures like, were done in few visits,

- Fluoride varnish for all teeth
- Pit and fissure sealants for all deciduous molars
- Glass ionomer restorations irt 55 53
- Pulpectomy irt 84, 85
- Extraction of 61, 63 and space maintainers were advised.

However for all the invasive procedures were planned to be done either under conscious sedation or general anesthesia subsequently but only after obtaining his physician's consent. The patient is kept under a close follow up.

DISCUSSION

Trisomy 21 or Down's syndrome is a genetic disorder caused by the presence of all or part of a third copy of chromosome 21. Like most of the trisomies, the incidence of trisomy 21 is highly correlated with maternal age, increasing from about 1/1500 live births for women 20 years of age to 1/30 for women 45 years of age and older as seen in the present case. Fig (2) (Harrison, 1950)

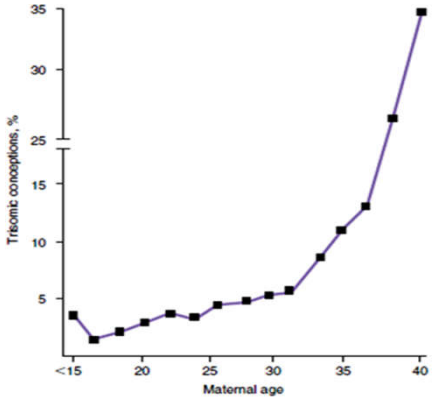


Fig. 2. An increase in the incidence of Trisomies with increasing maternal age

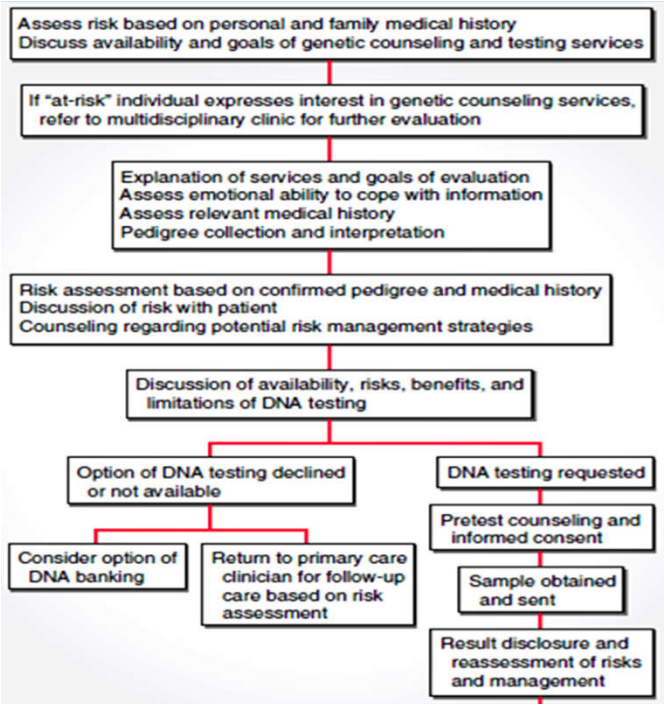
DS can be caused either by complete, mosaic or translocation type of trisomies, the most common cause being complete type wherein the complete extra copy of chromosome 21 is in all of the person's cells. The oral health of these subjects is greatly affected by their medical and physiologic characteristics, thus affecting their quality of life directly or indirectly (Ronald *et al.,* 1973). Several characteristics orodental features are seen in these patients. Individuals with DS present with about 35% to 55% of microdontia in both the primary and secondary dentition (Bell *et al.,* 2001, Kieser *et al.,* 2003). Clinical crowns are frequently conical, shorter, and smaller than normal (Townsend 1983, 1987). Both Japanese (Kumasaka *et al.,* 1997) and Brazilian studies (Acerbi *et al.,* 2001), found 60-63% of DS individuals had one or more missing teeth. In a detailed study, Russell and Kjaer (1995) studied 100 DS individuals and compared with Danish normal population, in their study missing teeth had a 10 times greater frequency in DS individuals than in general population and a higher frequency in males than in females as seen in the present case. A relatively high frequency of mal-alignment is also seen in both the deciduous and permanent dentition in individuals with DS. Some individuals having an open-mouth posture causes in-coordination of the lips, and cheeks in swallowing and speech. All these features were seen in the present case. Individuals with habitual mouth breathing tendencies are also more susceptible to periodontal disease. DS individuals have many common oral soft tissue manifestations like large and fissured tongue, cracked lips (Figure 1). The tongue is large (macroglossia) relative to the size of the oral cavity. Studies have shown that the tongue size of these individuals does not differ significantly from that of the general population (Ardran *et al.,* 1972). It is only the oral space, which is small (Guimaraes *et al.,* 2008). There can be marked fissuring of the

dorsum of the tongue, and because of poor muscular control, the tongue is often protruded, seen in our case.

Maxilla in DS individual is deficient in the development and the mandible is of normal size or slightly hypoplastic (Boyd *et al.,* 2004). DS patients have a narrow, high arched V-shaped palate, which is due to deficient development of the midface, affecting the length, height, and depth of the palate. Westerman *et al.* (1975) compared 40 DS individuals with 44 control subjects and concluded that the palatal dimension were narrower in width, shorter in depth, and lower in height and he concluded that the terms high arch and narrow palatal vault (Figure 1) were subjectively described and was only partly correct. These patients have a varied degree of intellectual impairment. Atlanto axial joint instability (AAI), common in DS patients, is characterized by hypermobility of C1-C2 joint due to laxity of alar ligament. The atlantoaxial instability has a prevalence of 22.5% in individuals with Down syndrome and the ligamentous hyperlaxity has a prevalence of 61.2%. Neck of DS individuals during dental treatment should be maintained in a relaxed position in order to avoid AAI from occurring (Burket's, 1994). DS can be differentiated from other trisomies like Edward and Patau syndromes which are trisomies of 13 and 18 chromosomes respectively. However the screening and diagnosing these trisomies remain the same with different clinical features. Screening can be done either prenatal or postnatal by Amniocentesis or umbilical cord blood sampling for karyotype determination and by chorionic villus sampling. Many complications are associated with DS, Alzheimer's disease, leukemia, heart defects, endocrine disorders and immune defects.

Dental management for down syndrome patient

Treatment objectives for patients with developmental disabilities like DS may need some adaptations and modifications but the goal must be to provide as comprehensive treatment as possible. It is a challenge for the physician and the pedodontist to do treatment and build a good level of trust in this population with delayed learning. Appointments must be scheduled early in the day as it is beneficial for both patient and operator is more rested. First appointment should be only for orientation and subsequent appointments may require a little more time than what is usually allowed. The patient's medical history should be obtained prior to the treatment as this allows for medical consultation if necessary before any treatment begins. Patient's caregiver must be encouraged to be present during the treatment. Children with DS should be educated in proper oral hygiene, occlusal sealants are recommended and topical fluoride therapy must be done periodically. Periodic oral prophylaxis and periodontal treatment must be done to maintain the oral hygiene and an antiseptic mouthwash can be advised. Flossing may be difficult to perform, hence mechanical toothbrushes can aid in maintaining a good oral hygiene. Due to AAI care should be taken during radiographic procedures, so as to limit the neck and head movements also the neck should be maintained in a relaxed position during dental treatment to avoid AAI from occurring. Functional orthodontic treatment such as palatal expansion with a removable appliance combined with Oromotor therapy can be done. DS individuals exhibit phonological-altered spoken communication with more unimodal gestural answer thus the dentist should find out from caregiver the patient's level of intellectual and functional abilities and communicate directly with DS individuals using short, clear instructions. An intensive preventive programme is recommended and should include: regular oral hygiene motivation, dietary counseling, topical fluoride and fissure sealants application. Periodontal treatment needs are higher in these patients. Modification of periodontal therapy can be done which involves non-surgical periodontal therapy adjunct with regular use of chemical plaque control agents, and frequent recall schedule in DS adults may be a way forward. As motor development is usually delayed in these children may lead to reduced manual dexterity. Parents and caregivers should be educated on the need to help with tooth brushing until the individual has acquired sufficient motor skills thus improving their oral hygiene. Most of the children are affectionate and cooperative for dental treatment; while some may require treatment under sedation or general anaesthesia like that in the present case, who was uncooperative. Children with DS exhibit AAI and extreme care is needed during intubation and orientation of the head by the paediatric dentist during provision of dental treatment under general anaesthesia. It is also difficult to do endotracheal intubations due to a large protruberant tongue, high arched palate, small mouth, short, broad neck, abnormal dentition, small maxilla & mandible, large tonsils. Due to laryngeal atresia, congenital sub-glottic stenosis; poses problems for even general anaesthesia, 2-3 mm endotracheal tubes are used. Genetic counseling must be given to married couple to prevent consanguineous marriages. The chart given below has the protocol to be followed Figure (3) (Burket's, 1994).



There is recent interest in the therapy for people with DS which has focused on pharmacological treatment to enhance cognition. The use of chromosome engineering to generate

new trisomic mouse models and large- scale studies of genotype–phenotype relationships in patients are likely to significantly contribute to the future understanding of DS. Hence to develop new therapeutic targets, it is necessary to determine the identity of genes that contribute to DS phenotypes and this requires a precise and standardized definition of phenotype. These measurements should be formulated into a standardized protocol that can be applied at multiple centres, to permit sufficiently large numbers of samples for meaningful analysis to be collected. This can be facilitated by a carefully designed and curated biobank of detailed phenotypic data alongside DNA and tissue samples from participating individuals (Kate *et al.,* 2009).

Conclusion

To conclude, DS individuals are basically a group of patients requiring special oral health care services. They have more orodental issues and are often affected with malocclusion. The dental care must be provided with special dental care services for this population, as they are unmet. However there are inadequate resources in many communities for DS subjects or their caretakers to upkeep their oral health is also less. DS care providers should acquire appropriate level of oral health awareness and communities and consider improving the accessibility of DS subjects to oral health care in order to assist maintenance of oral and overall health for this group of special need patients.

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