Dental Implications of Down Syndrome (DS): Review of the Oral and Dental Characteristics

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Abstract
A literature search was conducted to identify the key oral and dental manifestations of DS. These findings are discussed and used to suggest recommendations for treatment planning in DS patients for the practicing dental practitioner and also to help other medical professionals in understanding the oral health status of DS patients and the importance of liaison with dental professionals.

INTRODUCTION
DS or Trisomy 21 is a genetic disorder caused by a trisomy of an extra chromosome No.21 [1,2]. An abnormal segregation of chromosomes during cell division gives the affected individuals three instead of the normal two [3-5].

DS is the most common chromosomal condition diagnosed in the United States [5]. Approximately one out of every 700 infants born in the United States is diagnosed with DS [6]. Regionally, DS prevalence is around 1 in 554 in Saudi Arabia and 1 in 853 in Hyderabad in India [7,8]. Surprisingly, Dubai has scored the highest incidence in the region, one in every 319 live births among UAE nationals and one in every 602 live births among non-nationals are diagnosed with DS [9].

As the life expectancy among this population is increasing [10], schools, work and community settings are becoming the norm for DS persons [10,11]. The demand for dental care for this group with special needs is also increasing with this incidence trend and thus every practitioner should have a clear understanding of DS’s unique characteristics that would undoubtedly influence their dental care and treatment.

The aim of this paper was to identify the key oral and dental manifestations of DS and to suggest recommendations for treatment planning in DS patients for the practicing dental practitioner and also to help other medical professionals in understanding the oral health status of DS patients and the importance of liaison with dental professionals.
MALOCCLUSION

The prevalence of different malocclusion types has been investigated in different studies. Soares et al., found that Class III malocclusion is more common in DS children [12,13]. This Class III malocclusion is due to underdevelopment of the midface and not to prognathism of the mandible. The presence of Class III malocclusion has been confirmed by Jaber as well, where his DS study group showed higher prevalence of Class III malocclusion than the normal group [14].

In regards to posterior and anterior crossbite, Soares et al., found a 39% prevalence of posterior crossbite and 26% anterior crossbite [12]. Other studies reported a prevalence of 31% posterior crossbite and 33% anterior crossbite[12,15]. Almost similar results were found in a sample of DS in Sharjah city in the United Arab Emirates, where the prevalence of crossbite was 26% and 10% of open bite. In addition, high arched palate, fissured tongue and macroglossia have been reported to be more frequent in DS children than normal children[12,14,16,17].

DENTAL ANOMALIES

Anomalies of number, shape, structure and position of teeth are frequently observed in DS patients. Both primary and permanent dentition are affected and the incidence is approximately five times greater in DS children than in general population [12,18]. The most observed anomalies are hypodontia, delayed eruption, hypoplasia, supernumerary teeth, ectopic eruption, atypical patterns of eruption and abnormal dental morphology. Figures 1, 2 and 3 show respectively: transposition of upper canine, microdontia of upper second premolar and nipple appearance of the lower canine.

The prevalence of hypodontia is diverse in different ethnic groups [19]. Third molar agenesis was found to be around 4 times greater in DS than normal individuals. The latter is followed in decreasing order by agenesis of mandibular central incisors, maxillary lateral incisors, maxillary second premolars and mandibular second premolars [19,20]. The pattern of agenesis in DS is thought to be associated with peripheral nervous system abnormalities and abnormal cartilaginous tissue [19,20]. Hypodontia prevalence is reported to be around 60% in DS children [19,21,22]. Other anomalies described in the literature are macrodontia, microdontia, talon cusp, dens evaginatus, double teeth, amelogenesis imperfecta, dentinogenesis imperfecta, taurodontia, peg shaped teeth and impacted teeth [12,19]. The roots tend to be conical in DS individuals and this finding is significant when considering orthodontic tooth movement and periodontal disease [3].Tooth anatomy can affect the degree of root resorption as teeth with pipette shaped and blunt roots are significantly at greater risk of root resorption [23].

NON CARIOUS TOOTH WEAR

Bruxism

Bruxism is defined as parafunctional behaviour of the mandible, characterized by clenching or/and grinding of the teeth" [24,25]. It has been reported in the literature that bruxism prevalence is higher in children with cognitive impairment compared to normal children [24]. DS children have bruxism at a young age and usually it persists throughout life [12]. The factors that are thought to contribute to this phenomenon are that DS children have underdeveloped nervous system, malocclusion, chronic anxiety, temporomandibular joint dysfunction, hypotonicity and laxity of the supporting ligaments [12,26].

The discomfort of the malocclusion in DS children might unconsciously make them protrude their mandible to get a more comfortable position. This latter position traps the maxilla
behind and retards its growth furthermore. This mandibular protraction is also facilitated by the temporomandibular joint laxity [27,28]. The child might also clench or grind his teeth in an attempt to eliminate interferences in his/her occlusion and find a comfortable position [27,28]. Bruxism on the long term creates tooth wear facets, teeth fractures and overloading of the supporting tissues [24]. On the other hand, other studies found similar or less bruxism habits in DS compared to normal children and that could be a result of the variability in the diagnostic criteria of bruxism between studies [24].

**Erosion**

Tooth wear due to acidic and chemical assault to the teeth are commonly noticed in DS children [29]. This issue is related to the fact that 13.8% to 59% of DS children suffer from gastric dysfunctions like vomiting and gastroesophageal reflux [29-31]. A study by Bell [25], showed that dental erosion was significantly higher in DS individuals than the normal population which accounts to 67% compared to 34% in normal people.

The dentist should take a careful note of tooth wear in DS children and try to identify the aetiology in order to avoid the problems of dentinal hypersensitivity and dental destruction [29].

**ORAL DISEASES**

**Caries (Dental decay)**

As far as caries is concerned, the majority of the literature and researches describe a low prevalence of dental caries in DS children both in the primary and permanent dentition [12,26,32-34]. However, some studies reported similar caries rate between DS and normal control children [32,35,36], while others reported that DS children have more caries than healthy children [14,37]. The results are conflicting and this could be attributed to the inappropriate study designs, the sample number used, and not controlling covariates [38].

The literature attributes the reduced caries risk in DS individuals to several factors such as higher salivary pH [39], higher salivary bicarbonate levels which improves its buffering capacity [40], eruptive pattern (delayed eruption of teeth so they have less time to be exposed to cariogenic factors), bruxism (teeth are flatter and have reduced fissure depths so debris do not accumulate easily and the surfaces are self-cleansing), hypodontia (makes the dentition spaced) and microdontia (spaces are present between teeth and visual detection of caries is easier and earlier). Also, due to the nature of their complex medical condition, their parents tend to be more concerned about their dental health and seek dental advice earlier [12,26,32,41,42].

Regardless of the favourable factors mentioned above, the dentist should not underestimate the occurrence of dental caries in this group of children. DS children might have some dietary and oral hygiene habits that put them at a higher risk of developing gross caries [39]. When compared to normal children, DS children are more likely to bottle feed during sleep (50% compared to 12%), are on medications that contains sugar, have less help with their brushing and are weaned off bottle at an older age [43].

**Periodontal disease**

Gingivitis is an “inflammation of the gingiva in the absence of clinical attachment loss”. Clinically, it is noted as redness and oedema of the gingiva with bleeding upon probing. Gingivitis has no radiographic evidence of bone loss [44]. On the other hand, periodontitis is inflammation that involves the gingiva and the adjacent apparatus. Periodontitis is characterized by clinical attachment loss and loss of the adjacent supporting bone [44].

Periodontal disease in DS individuals has been first described by Nash, where she reported that 90% of DS patients exhibit some evidence of periodontal disease [45]. The sample she examined included children below 7 years and she suggested that the gonads hypofunction is the main reason, which is not accepted nowadays [45]. The occurrence of periodontal disease in DS patients is mostly due to defective immune system rather than poor dental hygiene on its own [33]. All of the longitudinal studies along with the cross sectional studies reported that the prevalence of periodontal disease in DS individuals is very high and can rapidly progress especially in the young age groups [10,12,45]. The prevalence has been reported between 90% and 96% in adults with DS [11,33]. The periodontal disease is also noted in the deciduous dentition [45].

The limited manual dexterity in DS children, lowered self-homecare and limited access to care all lead to poor oral hygiene and increased level of gingivitis [10]. Gingivitis can differ in DS children than healthy children. In an experimental gingivitis study, it was found that DS children developed rapid and more extensive gingivitis around deciduous teeth than normal control children. The amount of plaque between the two groups was similar [46]. Other studies tried to explain this and reported that there are no differences between the plaque composition in DS children and healthy children; however abnormalities in host defence particularly in leucocyte response may be the reason [47]. This pattern of gingivitis in DS has been also explained by the presence of defective connective tissue and altered vascularisation [47].

The greater concern about the periodontal disease in DS individuals is the progressive pattern of the disease. Children with the syndrome can present with marginal gingivitis, gingival recession, advanced periodontitis and pocket formation. Brown and Cunningham found that 36% of DS children had pocket formation below the age of 6 years [45]. They can also experience acute necrotizing ulcerative periodontal disease more frequently.
The tongue is forced to form an oral seal which will affect the bite and an incomplete lip closure. Lack of lip seal is also caused by maxillary teeth will erupt in an edge to edge relationship or with malocclusion [12]. The maxilla will be underdeveloped and the unfavourably influence the shape of the maxilla leading to [27]. This unfavourably muscular weakness of the tongue will and in a low position in the mouth (relative macroglossia) large as a result of muscle weakness which makes it sit anteriorly and in a low position in the mouth (relative macroglossia) [45].

There are many researches that highlighted the abnormalities in the DS immune system including the non-specific defence mechanism, the cellular and the humoral immune systems [45]. Several defects have been reported in advanced periodontal destructions in DS such as diminished chemotaxis of neutrophils, decreased phagocytic ability and shortened half-life of the neutrophils [10]. The polymorphonuclear leucocytes (PMN) activity towards aggregatibacter actinomycetemcomitans (AA) is reduced in DS individuals compared to age matched controls [10]. The PMN defect in DS is a qualitative type, where there bactericidal function fails and the neutrophil adhesiveness to bacteria is reduced [45]. An integral feature of DS immune system is defective T-cell maturation, low level of immunoglobulins IgM and altered function of B-cell lymphocyte [45].

The amount of periodontal pathogens in DS individuals has been found to be higher than patients with other mental challenges. Higher amounts of P. gingivalis, motile organism, Tannerella forsythia and spirochetes have been reported in different studies. Viruses have also been reported to co-exist with the periodontal pathogens in some DS cases such as Epstein-Barr virus, human cytomegalovirus and herpes virus [10].

Treatment of periodontal disease in DS children can be very challenging and the family plays an important role in the treatment. DS children have compromised capacity in performing oral hygiene and parents should get involved and supervise them [10]. Despite treatment, some cases can show severe destructive pattern. A longitudinal study was done by Barr-Agholme to measure the progression of periodontal disease[48]. He found that most of the patients showed increased bone loss from 35% to 74% particularly in the mandibular incisors.

**TONGUE ABNORMALITIES**

**Macroglossia**

The orofacial characteristics are greatly influenced by facial muscle hypotonia. The tongue in particular looks abnormally large as a result of muscle weakness which makes it sit anteriorly and in a low position in the mouth (relative macroglossia) [27]. This unfavourably muscular weakness of the tongue will unfavourably influence the shape of the maxilla leading to malocclusion [12]. The maxilla will be underdeveloped and the maxillary teeth will erupt in an edge to edge relationship or with a reverse overjet. Both the lingual tongue posture and facial muscle hypotonia result in imbalance in the muscular forces between the lip and the tongue, which develops an anterior open bite and an incomplete lip closure. Lack of lip seal is also caused by the hypotonic lip muscules [27]. As a result of this open bite, the tongue is forced to form an oral seal which will affect the swallowing action. The swallowing action can be compromised further if the tongue is used to stabilise the mandible against the maxilla [27]. Muscle hypotonicity can also cause joint hyper-flexibility and saliva drooling at the labial commissure. The latter will lead to angular cheilitis, aphthous ulcers, cracking, and candidiasis [12].

**Fissured tongue**

Another tongue abnormality that has been reported in DS children is fissured tongue [3,12,14]. Fissured tongue is a non-pathological variation of the normal tongue, where the dorsal surface of the tongue is altered by the presence of a central groove and several clefts resembling veins of a leaf [49]. Microscopically, the main feature is the presence of various sizes of papillae and more inflammatory cells than in normal tongue [49]. This condition is usually asymptomatic and can sometimes be associated with geographic tongue. The exact aetiology is unknown and a polygenic mode of inheritance is suspected. Fissured tongue is noted on routine dental examination. These deep fissures can act as bacterial reservoir and cause glossitis [50]. A DS individual also may have the ability to extend her/his tongue and touch her/his tip of the nose, which is known as Gorlin sign as shown in (Figure 4).

**ACCESS TO DENTAL CARE**

Today, no statement of reasons is needed to say that optimal oral health is an essential prerequisite for good general health. Oral health optimises self-esteem, nutrition communication and quality of life [51]. Although, the oral health has been improved over the decades, some groups might still experience suboptimal oral care.

Children with special needs have specific intellectual, physical and psychological problems and should get special oral care in the dental office [52]. Special need children may have jeopardized oral health because of their medical issues, use of medications, craniofacial defects, teeth anomalies, enamel abnormalities and difficulty in practicing the routine oral hygiene measures [51].

The insufficiency in these children’s oral care is not only because of their physical and intellectual challenges, but could also be due to barriers to proper oral healthcare [51]. These barriers are either environmental or non-environmental. The environmental barriers are focused on the oral care delivery system such as insurance, financial aspects, finding a dentist that will accept to treat the disabled child. On the other hand, the non-environmental barriers are those that originate in the special need individual himself such as anxiety, dental phobia, medical conditions that complicated his/her dental treatment [51].

A study was conducted to compare oral health care utilization between special needs children and healthy ones during a 7 year period in Belgium. This study found that 50% of the special needs children had only one dental visit in four or more of the seven observation years. Most of the visits were emergency visits. On the other hand, the healthy group had dental visits for radiographs, restorations, orthodontic assessments and treatments. The same study stated that there were very low rate of attendance among the special needs children group and preventive oral health care was not frequently received [51]. Another study which was
conducted in Canada found that children with DS received less restorative work and more extractions compared to their healthy siblings [38].

Looking at another study in India, where they compared dental care between DS children and their siblings through a questionnaire filled by their parents, the study found that DS children received different oral care than their siblings. DS children were less likely to visit a dentist yearly, less likely to receive restorative treatment and caries prevention and less likely to have dental extractions [53]. The author concluded that it is a cumulative neglect and mostly parental neglect of their children’s basic health measures. It also reflects lack in the overall scheme of health management to this disadvantaged group of children [53].

Children with DS vary and often they lack cooperation or have neuromuscular problems, craniofacial deformities and joint laxity which make the routine oral hygiene measures difficult [52]. Several studies have found that DS children exhibit poor oral hygiene compared to normal children [52,54]. In the UAE, a study of DS children showed that they have poor oral hygiene, higher occurrence of periodontal disease and dental caries compared to normal matched children [14]. Parents and caregivers should be encouraged to assist their children to accomplish acceptable oral hygiene measures. Therefore, the dentist should educate the parents as part of the prevention plan for DS children [52].

CONCLUSION

Down syndrome individuals are special needs patients with several medical and orofacial abnormalities. The provision of dental care to these individuals presents unique challenges to the dental staff. Therefore, dentists dealing with them should possess thorough knowledge of the unusual medical and orofacial abnormalities and their implications and should liaise with different medical specialists in order to formulate safe and effective dental preventive and treatment plans. Prevention of dental diseases in DS individual is of paramount importance especially for those who are severely or profoundly medically compromised.

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REFERENCES


