

Down Syndrome-An Insight to Dental Aspect- Case Report

Ambarkova Vesna^{1*} and Manu Batra²

¹*Faculty of Dental Medicine, Department of Paediatric and Preventive Dentistry, University St. Cyril and Methodius, Skopje, Republic of Macedonia*

²*Senior Lecturer, Department of Public Health Dentistry, Surendera Dental College and Research Institute, Sri Ganganagar, Rajasthan, India*

***Corresponding Author:** Ambarkova Vesna, Faculty of Dental Medicine, Department of Paediatric and Preventive Dentistry, University St. Cyril and Methodius, Skopje, Republic of Macedonia.

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Abstract

Down's syndrome is the most common genetic cause of significant intellectual disability in the human population. It caused by the presence of all or a part of a third copy of chromosome 21. This article reports a case of Down syndrome in a 15year old male patient who reported to Department for Preventive and Pediatric Dentistry, University Dental Clinic Center Ss. Pantelejmon, Skopje.

Keywords: Down Syndrome; Mental Retardation; Chromosome 21 Trisomy

Introduction

Down's syndrome is defined as a genetic disorder caused by the presence of all or a part of a third copy of chromosome 21. It was named after John Langdon Down, the British physician who described the syndrome in 1866. This syndrome is also known as Trisomy 21, named by Dr. Jecome Lejeune as a common chromosomal abnormality [1]. The incidence of Down syndrome rises with increasing maternal age. The features of Down syndrome can range from mild to severe. Usually, mental development and physical development are slower in people with Down syndrome [2,3].

In these patients, there is a strong predisposition to cardiovascular disease, seizures [4], leukemia [5,6], infections with hepatitis B virus (especially within institutionalized men) [7], upper respiratory tract infections [8], Alzheimer's disease [8], obesity [9], thyroid diseases [10], cardiac anomalies [11] and obstructive sleep apnea [12-14]. Disruption of the proteostasis network and accumulation of misfolded proteins occur as a result of an abnormality in the number of chromosome 21 [15]. Errors in protein homeostasis could contribute to the observed pathology and decreased cell viability in children with Down syndrome [16].

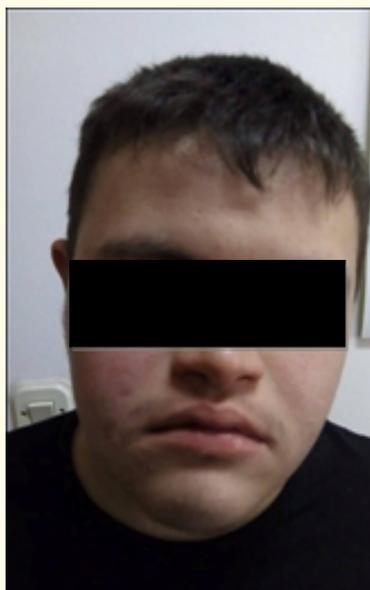
Case Report

A 15-year boy reported to the Department for preventive and pediatric dentistry, University Dental Clinic Center-Skopje with a complaint of toothache in right lower first molar tooth since 4 days. The family history revealed that he has two siblings, two sisters (9 and 18 year old) with no syndromic features.

On general examination, patient is short statured and mentally challenged. On extra oral examination patient had saddle nose deformity, midface hypoplasia with retruded maxila and protruded mandible. Intraorally, high arch palate was present. Macroglossia of the tongue was present.

A detailed clinical examination revealed typical features of Down syndrome. The orthopantomogram shows the presence of a periapical lesion in 46. The patient was not cooperative therefore endodontic treatment was ruled out and the extraction of the tooth was indicated.

An opening of the first right lower molar was performed using a high speed dental turbine with diamond bur drill in the enamel and with a Slow Speed Contra Angle handpiece and new steel bur in the dentine area. We applied the Solution Chlumsky (Sitisan, product from manufactory Galenika, Beograd) to calm the pain of the tooth in the patient. We sent him to the Oral Surgery Clinic to extract the tooth (46).



Frontal View



Left Profile View



Right profile view



Tongue



Orthopantomograph view

Discussion

The signs and symptoms of Down's syndrome are characterized by neotenzation of brain and bodies. Management strategies such as Early childhood intervention, screening from common problems, medical treatment when indicated, a conducive family environment and vocational training can improve the overall development of children with Down's syndrome.

The changes in body of the children with Down syndrome are manifest through the short stature, short legs and arms, slow skeletal maturation, poor muscle development, always with the presence of obesity, poorly developed male genitalia, short and wide neck, dry and rough skin, straight and smooth hair. The changes in the head and face are characterized by brachycephaly, microcephaly, undeveloped vertex bone (os parietale) of the skull that gives the appearance as if the back of the skull is flattened. Also nasal bones are hypoplastic, with flat nasal bridge. The combination of minor and major abnormalities characterize this syndrome that give typical phenotypic appearance of the head and face: hypertelorism, small and oblique eyes, spots on the iris, strabismus in 30% of patients, deformed and small ears. The limbs are short, the typical appearance of the hands with ruffle and short fingers and transverse crease on the palms present in about 80% of cases [17].

Open mouth with protrusion of the hypotonic and fissured tongue, macroglossia, hypoplastic upper jaw, short and hard palate, pseudoprognathic bite, delay eruption of the teeth [17], persistence of some deciduous tooth up to 15 years, the order of eruption of teeth is disturbed [18], hypodontia, microdontia, atypical form of dental crowns and taurodontism [19]. Also patients with Down syndrome can present periodontal disease, premature tooth loss, reduced salivary flow, crowding of teeth in both arches, and decreased occlusal vertical dimension. Increased frequency of periodontal disease, reduced incidence of dental caries [20], often spilling of saliva from the mouth. In these patients, cleft lip, cleft lip and palate or palate only occurs 3 times more often than in the general population. The findings by van der Linden MS., *et al.* showed that dental development in children with Down syndrome is similar to the development of control children and that a relationship exists between hypodontia and dental development [21,22].

For patients with mild mental impairment, Altintas NY, *et al.* in their case have suggested implant-retained overdentures with Locator attachment system [23].

Before starting of the implementation of the orthodontic treatment within the patients with Down syndrome, it is necessary to restore all decay teeth. If the patient is unable for clinical work, orthodontic treatment can be initiated with a mobile device [24], which usually carried extension of the upper jaw and, if necessary, "pull" of the front teeth due to the presence of the pseudoprognathia [25]. Treatment can be continued with a fixed appliance that compensates dental chains, regulates the placement of the teeth in the dental chains and adjust dental bite [26]. Miyazaki H., *et al.* describe orthodontic treatment in a patient with Down's syndrome with unilateral cleft lip and alveolus. After surgical intervention and further mental and physical growth of the patient, the orthodontic treatment was indicated with multi-bracket. Reverse occlusion was corrected by labial inclination of the incisors [27].

Fakhruddin KS., *et al.* from their clinical study involving 22 children with Down syndrome concluded that routine psychological (Tell-Show-Do) intervention along with visual distraction using video eyewear and use of CDS-IS (computerized delivery system-intrasulcular) system for anesthetic delivery is effective behavior management technique during invasive dental treatment [28].

Considering the great emotional, psychological and material goods that require such member of a family and the modest effects of the expensive therapy, enormous efforts to the prenatal diagnosis are invested nowadays [29-32]. Prenatal diagnosis of a mother who previously gave birth to a child with trisomy, and in women who give birth after the age of 35 years, significantly reduces the number of children born with the disease. However most of children with Down syndrome remain undetected prenatally.

Resently it's believed that with the analysis of certain ultrasonic parameters associated with the combination of determining α fetal-protein, estriol and β HCG (beta-chorionic gonadotropin) in the mother's blood at the 14th week of pregnancy can be assign degrees of urgency to certain group of pregnant women with the increased risk of delivering a child with trisomy 21. Kochova and its associates [33] considered that this group should undergo prenatal diagnosis by amniocentesis. In this manner 75 - 80% of pregnancies with a Down syndrome fetus can be detected.

Adults with disabilities in Macedonia are employed at a significantly lower rate than adults without disabilities. The situation of the unemployed persons with disabilities is also similar in other countries of the world, even in the United States [34].

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