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List of abbreviations

A.a: Actinobacillus actinomycetem commitans.

AAC: Augmentive and alternative communications.

APF: Acidulated phosphofluoride gel.

CHD: Congenital heart diseases.

DS: Down Syndrome.

DMFT: Decayed, missing, filled permanent teeth .

deft: decayed, missing, filled primary teeth.

GERD: Gastroesophageal reflux disease.

GI: Gingival index.

NaF: Sodium fluoride.

S.mutans: Streptococcus mutans.

TiF4: Titanium fluoride.

TMJ: Temporomandidibular joint.

TW: Tooth wear.

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Marwa Abdelhak Ibrahim Moustafa

Dedication

To my beloved Father and Mother and my whole Family who are always there for me,

I will be always and forever indebted to you.

*Introductio*n

Down syndrome (**DS**) 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. Trisomy 21 is the most common chromosomal abnormality in humans. It is typically associated with a delay in cognitive ability and physical growth, as well as a particular set of facial characteristics [1].

The birth incidence of DS varies between countries and is generally cited as being between 1 in 600 and 1 in 1000 live births[2]. In Egypt, there is an estimated risk of 2285 DS births annually and 1.6 million births per year.[3].

Individuals with DS have a higher risk for many conditions. The medical consequences of the extra genetic material in DS are highly variable. It may affect the function of any organ system or bodily process, and can contribute to a shorter life expectancy for people with DS[4].

Systemically, DS is characterized by decelerated maturation (neoteny) of the brain and body, intellectual disability, congenital cardiac defects, midfacial hypoplasia and brachycephaly**.** Several oral manifestations have been reported in DS such as delayed eruption of teeth, abnormal pattern of eruption, congenital missing teeth and periodontal diseases[4-6].

A number of reports have outlined the impact that dental caries and periodontitis have on the dentitions of individuals with DS[7-9]. However, an

important oral condition that has largely been ignored in these individuals is pathological tooth wear. The process of tooth wear has a multifactorial etiology which is subdivided into attrition, erosion, abrasion and abfraction. It is likely that the heavy levels of tooth wear in DS are associated with tooth grinding and an acidic oral environment[7] .

Therefore, the aim of the present study was to assess the oral health status in a group of children with DS, characterize the diagnosis and etiological factors associated with tooth wear and assess the effect of different management strategies in its progression.

Review of literature

Down syndrome

Down syndrome (**DS**) was named after John Langdon Down, the British physician who described the syndrome in 1866. The condition was clinically described earlier by Jean Etienne Dominique Esquirol in 1838 and Edouard Seguin in 1844. DS was identified as a chromosome 21 trisomy by Dr. Jérôme Lejeune in 1959. DS can be identified in a baby at birth or before birth by prenatal screening [1].

Etiology:

A typical human karyotype is designated as 46,XX or 46,XY, indicating 46 chromosomes with an XX arrangement typical of females and 46 chromosomes with an XY arrangement typical of males [10].

There are 3 types of chromosomal abnormalities that can cause DS[10]:

1- Regular trisomy 21 or nondisjunction.

2- Translocation; where part of chromosome 21 is attached to another chromosome.

3- Mosaic type; where a mixture of trisomy 21 cells and regular cells exists in the body.

1- Trisomy 21:

Trisomy 21 (47,XX,+21) is caused by a meiotic nondisjunction event. With nondisjunction, a gamete (*i.e.*, a sperm or egg cell) is produced with an extra copy of chromosome 21; the gamete thus has 24 chromosomes. When combined with a normal gamete from the other parent, the embryo now has 47 chromosomes, with three copies of chromosome 21. Trisomy 21 is the cause of approximately 95% of observed DS cases, with 88% coming from nondisjunction in the maternal gamete and 8% coming from nondisjunction in the paternal gamete [10] .

2-Robertsonian translocation:

The extra chromosome 21 material that causes DS may be due to a Robertsonian translocation in the karyotype of one of the parents. In this case, the long arm of chromosome 21 is attached to another chromosome, often chromosome 14. A person with such a translocation is phenotypically normal. During reproduction, normal disjunctions have a significant chance of creating a gamete with an extra chromosome 21, producing a child with DS. Translocation DS is often referred to as *familial DS*. It is the cause of 2–3% of observed cases of DS. It does not show the maternal age effect, and is just as likely to have come from fathers as mothers [11].

3- Mosaic type:

In this type, some of the cells in the body are normal and other cells have trisomy 21, this is called mosaic DS. It accounts for 1-2% of DS cases [11].

Epidemiology:

The Center of disease control estimates that about 1 of every 691 babies born in the United States each year is born with DS. Each year about 6,000 babies in the United States are born with this condition. Approximately 95% of these are trisomy 21 [12].

In Egypt, there is an estimated risk of 2285 DS births annually and 1.6 million births per year [3].

In 1998, Heuther et al. [13], assessed the maternal age as a risk factor for DS. The researchers reported that at maternal age 20 to 24, the probability was one in 1562; at age 35 to 39, the probability was one in 214, and above the age 45 the probability was one in 19. The authors also reported that although the probability increases with maternal age, 80% of children with DS were born to women under the age of 35, reflecting the overall fertility of that age group.

Clinical findings:

I. Systemic findings:

1-Physical characteristics:

DS is characterized by decelerated maturation (neoteny) of the brain and body. Individuals with DS may have some or all of the following physical characteristics or dysmorphic features as microgenia (abnormally small chin), oblique eye fissures on the inner corner of the eyes, muscle hypotonia (poor muscle tone), a flat nasal bridge, a single palmar crease, a flat and broad face, as well as a short neck. Other features that can be seen in