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Down syndrome-Trisomy 21: Oral and systemic considerations for the dental approach

Cristhiane Olívia Ferreira do Amaral [1, *], Rodrigo Bueno Badan [2], Carla Souza Andrade [2], Mariana Olívia Ferreira do Amaral [3] and Fabiana Gouveia Straioto [1]

[1] *Faculty of Dentistry, University of Western Sao Paulo, Presidente Prudente, Sao Paulo, Brazil.*
[2] *Special Program for Scientific Initiation (PEIC/SPSI), University of Western Sao Paulo, Presidente Prudente, Sao Paulo, Brazil.*
[3] *Medical School, University of Western Sao Paulo, Presidente Prudente, Sao Paulo, Brazil*

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Abstract

Objective: The physical, systemic and stomatological characteristics of patients with Down Syndrome (DS) were verified, for a better integration with the dentist and other health professionals.

Methodology: A search was carried out in the main databases and articles that were read in full and according to the objective, published as original research, case reports and literature review articles, 56 were **included.** Individuals with DS have unmistakable characteristics such as short stature and low-set ears, in addition to systemic impairments. On the stomatognathic system, they have injuries mainly in relation to malocclusion, dental caries and periodontics.

Discussion: The search for an oral health professional is late and the quality of life is negatively affected by changes that are often already aggravated. Dental surgeons should be more prepared to manage the care of these patients, and public policies focused on prevention should be addressed and are suggested.

Conclusion: According to the literature found, the physical and systemic characteristics of patients with Down Syndrome are easily identifiable by the dentist through a clinical look and anamnesis. In addition, oral health is more affected by dental caries, periodontal disease and malocclusion and professionals should be better trained to provide care with a focus on prevention.

Keywords: Down Syndrome; Trisomy 21; Oral Health; Dentist-Patient Relations

1. Introduction

Down Syndrome (DS) is a set of signs and symptoms [1] characterized by the presence of an additional chromosome 21 causing a condition known as trisomy 21 [2-8]. It can occur in three ways: free (error in the division during meiosis – approximately 95% of cases), Robertsonian translocation (the fusion of chromosomes 13, 14, 15 with 21 occurs) and mosaicism (presents normal and trisomic cells at the same time). In 1866, John Langdon Down was the first to make a complete description of this syndrome [1,5], but it was only in 1959 that J. Lejeune, M. Gauthier and R. Turpin identified its genetic cause [9].

Individuals with DS have an unmistakable phenotype, with a flattened face, oblique eyes with an epicanthal fold, low insertion of the ears, muscle hypotonia, tongue protrusion, short fingers, ligament laxity and variable cognitive deficits,

* Corresponding author: Cristhiane Olívia Ferreira Do Amaral

there are also underlying pathologies, such as heart disease and vision disorders. And effects that are deleterious, such as increased susceptibility to respiratory diseases and low immunity [10-13]. In addition, most have dental and stomatognathic disorders, and follow-up with an oral health professional is essential [14].

Among the changes and disturbances are: mouth breathing; xerostomia; atretic maxilla ; mandibular protrusion; atresic and deep palate; cracked lips and tongue; bifid uvula; altered papillary, marginal and inserted gingiva; macroglossia ; temporomandibular disorder; malocclusion, the most common being the Class III type, unilateral or bilateral crossbites. Alterations of dental structures are also observed, such as agenesis, late tooth eruption, taurodontia, microdontia, hypodontia, hyperdontia, enamel hypoplasia [15,16]. There is a higher occurrence of premature periodontal disease, tooth exfoliation, presence of calculus, gingival bleeding, candidiasis, cheilitis, acute necrotizing ulcerative gingivitis, mucosal ulcers. These characteristics are capable of generating impacts on mastication, articulation, swallowing and speech [2,3,17-18]. A recent study suggests that these individuals have a lower salivary density of *Streptococcus mutans,* but this cannot be related to a lower caries experience [3]. However, another study suggests that these individuals have a high rate of caries due to dental hygiene and cariogenic diet [16].

However, studies indicate that there is a low demand for dental offices by individuals with DS. The role of the dentist is fundamental for maintaining their oral health, as they tend to have poor hygiene and a cariogenic diet, socioeconomic factors, the limited knowledge of parents or guardians about the importance of dental care and the lack of dental care with specific programs for this population. It is necessary to use differentiated management techniques and special attention during the consultation, taking care with the type of medication used by the patient [16].

Therefore, it is important that the dental surgeon has knowledge about DS and especially about its systemic and oral implications, which require specific skills from this professional to deal with these patients.

The objective of this literature review was to verify the physical, systemic and stomatological characteristics of patients with Down Syndrome, for a better integration with the dentist and other health professionals.

2. Methodology

The present study was developed through the documental analysis of the bibliographical production obtained in the following databases: Virtual Health Library (VHL) – Bireme, LILACS, IBECS, MEDLINE and BBO – PubMed, SciELO, Brazilian Digital Library of Theses and Dissertations (BDTD) and in the Periodicals Capes database. Among the search strategies used, the following keywords were included: “Down Syndrome”, “Trisomy 21”, “Oral health”, “Behavior and Behavioral Mechanisms” and “Medical records”. The selection criteria were: articles according to the objective of the present study, in Portuguese, English and Spanish. Published articles were included, such as: original research, case reports and literature review articles and research that addressed themes inherent to Medicine, systemic condition, Dentistry and oral health of Down Syndrome, with full availability of the text in digital media (open access). After analyzing the studies, 56 references between were selected as the basis for the development of the literature review.

3. Literature review

Among the causes of Down Syndrome, advanced maternal age has been identified as the main one, due to the formation of gametes during the intrauterine phase and their interruption in meiosis I, causing these oocytes to age along with the woman, as they will only mature from puberty [1]. This aging would be capable of destroying chromosomal fibers and deteriorating the centromere, causing an inability to separate chromosomes during anaphase I of meiosis [5]. Another cause may be the lack of segregation in gametogenesis due to advanced paternal age (greater than 55 years). Inadequate consumption of alcohol, cigarettes, chemical substances, oral contraceptives, genetic inheritance, history of abortions and environmental agents, for example, radiation, are other causes that can cause genetic errors [1].

The physical characteristics of this syndrome are: short stature, shortening of the extremities (hands, feet, fingers, nose and ears), single transverse palmar crease, feet with wide space between the first and second fingers with a deep groove close to the plantar surface, genitals usually poorly developed, brachycephaly , wide and short neck, almond-shaped eyes relatively far apart, slanted palpebral fissure, absence of lip seal, narrow nasal bridge and nasopharynx, low-set ears, protruding cheeks, adenotonsillar hypertrophy, thin and sparse hair . It should be taken into account that there may be differences between the characteristics according to the racial group to which the individual belongs (Sudanese or Caucasian). They generally have a docile, sweet, affectionate and happy temperament [1,2,16,17,19-21].

These individuals may also have generalized muscle hypotonia, congenital heart disease, pulmonary hypertension, obstructive sleep apnea, hearing and ophthalmological impairments, thyroid disorders, 75% of which are hypothyroidism and 6% hyperthyroidism, diabetes, obesity, motor disorders, celiac disease, changes in cervical spine, neurological problems with greater language impairment and cognitive delays, 26% may have epilepsy and premature aging in these individuals may cause Alzheimer's disease in 25% of cases. It is also possible to observe the presence of gastrointestinal disorders, such as necrotizing enterocolitis, which occurs in 16% of newborns. Hypovitaminosis D and low bone mineral density can lead to osteoporosis in adults. Changes in the immune system lead to a higher prevalence of infections such as pneumonia and sepsis. As for the reproductive system, women are considered fertile and can transmit this syndrome to 50% of their offspring [1-4,6-9,16, 17,22-30].

Regarding oral problems, malocclusion, periodontal disease and dental caries are most prevalent, according to a recent literature review including observational studies [16], which is why these three were chosen for a more complete description in this study.

Individuals with DS have vertical and transverse changes, they are: mandibular protrusion (Class III), anterior open bite and anterior or posterior crossbite and dental crowding, this occurs due to a combination of factors, which are changes in the tongue (macroglossia)., facial and dental deformities, mouth breathing, among others. These alterations have repercussions on swallowing, speech and mastication problems, affecting the quality of life, therefore, some procedures may be indicated in orthodontics, such as rapid maxillary expansion with a palatal plate, palatine disjunction and even a fixed device for assertive alignment of the intercuspation dental. After these procedures, especially with maxillary expansion in the first years of life, there is an improvement in snoring, mouth breathing, drooling habits, articulation in words, muscle and lingual strengthening, and facial aesthetics, in addition to better airway clearance. upper airways [2,20,31-37].

Periodontal problems develop rapidly at an early age, this happens due to systemic and immunological factors (dysfunction of neutrophils and T lymphocytes, increase in inflammatory mediators and hyper innervation of the gum), in addition to motor and cognitive limitations, causing lack of manual ability to oral hygiene combined with mouth breathing, dental morphology, which can easily lead to gingivitis and then to periodontitis, also these individuals, when they have hyposalivation , are at greater risk of triggering severe periodontal disease. Parents and teachers need to be aware and seek information about effective approaches to dealing with oral hygiene with a professional. Individuals with DS can be trained when the impairment is mild or moderate [38-40].

Like the periodontium, the dental structure can be compromised due to oral hygiene, leading to dental caries, as there is a strong association with the fact that these individuals have less salivary flow and less buffering capacity [41], a study proved that electric toothbrushes and manuals are similar for biofilm removal [42], but another proved that the headphone technique aided by an electric brush is better than the manual technique to overcome the lack of motor dexterity [43].

Specific cases of diseases such as diabetes, celiac disease and leukemia can result in, respectively: severe periodontitis; tooth enamel defects and aphthous stomatitis; hemorrhagic gingival hyperplasia and ulcerations, which requires attention from the dentist [17].

4. Discussion

Due to the fact that parents and guardians of individuals with DS from an early age have to worry about medical problems that require ongoing treatment, oral health tends to be left aside and is seen as a low priority, despite pain, infections, problems chewing and speech, the search for a dental professional often happens when conditions are severely advanced [44-47].

In addition, the number of professionals who are trained to serve this public is extremely low, both in the public and private sectors, making it difficult for people with disabilities to find dental care, which is one of the reasons for the late demand for care with the oral health [45,46,48].

Regarding oral alterations, dental caries, periodontal disease and untreated malocclusion negatively affect the quality of life of children and adolescents with the syndrome, and some factors can be attributed to socioeconomic conditions, limited knowledge of parents and little access to dental care, which is why there is a lack of health promotion programs on the importance of prevention and the best ways to carry out hygiene and early diagnosis [16,49].

Unsatisfactory oral hygiene is considered one of the main problems, as procedures such as brushing and flossing to remove plaque on the tooth surface become a challenge. The physical and behavioral characteristics of DS can make cleaning difficult for parents and guardians, in addition, they have limited autonomy due to motor dysfunctions. Studies emphasize targeted public policies that generate a positive influence between the association of home plaque control and periodic visits to the health professional [50-52].

Understanding these facts, we move on to the following question: Lack of knowledge, negligence and unpreparedness of dentists for the clinical care of patients with DS. A study proved that most dentists do not have difficulty identifying, but they do not manage enough to perform the service, authors suggest that most did not have sufficient preparation during graduation. In addition, a meticulous anamnesis must be performed, checking medications that may interfere with dental treatment, referring to the doctor when necessary and even observing possible abuse [53,54].

The behavior of the dental surgeon must emit confidence, this is decisive in the attitude towards management. The consultation time should be short and calculated, not exceeding 30 minutes and the approach should be multidisciplinary together with the family, offering better conditions in terms of quality of life. Care for this patient must be in conjunction with diagnosis, therapeutic goals, reassessing and monitoring progress [55].

The focus on prevention is essential, and the oral health of patients with DS should be seen as a public health problem, currently there is a greater tendency for this population to be included in the job market, which further encourages the promotion of quality of life through federal laws and projects, allowing access to these patients at all levels of public and private health care [56].

5. Conclusion

According to the literature found, the physical and systemic characteristics of patients with Down Syndrome are easily identifiable by the dentist through a clinical look and anamnesis. In addition, oral health is more affected by dental caries, periodontal disease and malocclusion and professionals should be better trained to provide care with a focus on prevention.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

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