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# Down syndrome and dental aspects

Síndrome down y aspectos dentales Síndrome de down e aspectos odontológicos

## ABSTRACT

Objective: To review articles available in scientific journals that portrayed the dental aspects of patients with Down Syndrome. Methods: This is a bibliographic review study carried out in 2020, the time limit established for the research was between 2008 to 2018, eligible for the bases of the Brazilian Digital Library of Theses and Dissertations (BDTD), Coordination for the Improvement of Level Personnel Superior (CAPES) and Google Scholar. Conclusion: From the information obtained, we can conclude that supervised oral hygiene is essential in these individuals, due to motor limitations and the high prevalence of periodontal diseases. The dental surgeon must be prepared to assist patients with special needs through technical training and cultivation of human values, minimizing the various problems that these patients are already conditioned to face due to their disability. **DESCRIPTORS:** Down syndrome; Dentistry; Oral manifestations.

#### RESUMEN

Objetivo: Revisar artículos disponibles en revistas científicas que retratan los aspectos dentales de pacientes con Síndrome de Down. Métodos: Se trata de un estudio de revisión bibliográfica realizado en 2020, el tiempo límite establecido para la investigación fue entre 2008 a 2018, elegible para las bases de la Biblioteca Digital Brasileña de Tesis y Disertaciones (BDTD), Coordinación para el Perfeccionamiento del Personal de Nivel. Superior (CAPES) y Google Scholar. Conclusión: De la información obtenida se puede concluir que la higiene bucal supervisada es fundamental en estos individuos, debido a las limitaciones motoras y la alta prevalencia de enfermedades periodontales. El cirujano dentista debe estar preparado para atender a los pacientes con necesidades especiales mediante la formación técnica y el cultivo de los valores humanos, minimizando los diversos problemas a los que estos pacientes ya están condicionados a afrontar por su discapacidad.

**DESCRIPTORES:** Síndrome de Down; Odontología; Manifestaciones orales.

#### RESUMO

Objetivo: Revisar artigos disponíveis em periódicos científicos que retratassem os aspectos odontológicos de pacientes com Síndrome de Down. Métodos: Trata-se de um estudo de revisão bibliográfica realizado em 2020, o limite temporal estabelecido para a pesquisa foi entre 2008 a 2018, elegíveis as bases da Biblioteca Digital Brasileira de Teses e Dissertações (BDTD), Coordenação de Aperfeiçoamento de Pessoal de Nível Superior (CAPES) e Google Acadêmico. Conclusão: A partir das informações obtidas podemos concluir que A higiene bucal supervisionada é fundamental nesses indivíduos, devido às limitações motoras e à alta prevalência de doenças periodontais. O cirurgião-dentista deve estar preparado para o atendimento de pacientes com necessidades especiais por meio da capacitação técnica e cultivo de valores humanos, minimizando os diversos problemas que esses pacientes já estão condicionados a enfrentar devido ao seu estado de deficiência.

DESCRITORES: Síndrome de Down; Odontologia; Manifestações bucais.

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#### INTRODUCTION

Down syndrome was first described by the English physician John Longden Hayden Down, in 1866, and at that time it was also called Mongolian idiocy. However, it was only in 1959 that Down Syndrome was shown to result from the presence of an extra chromosome in pair 21, also being called trisomy of chromosome 21.<sup>1,2</sup>

Down's syndrome represents the most common chromosomal anomaly of the human species. Data show that Down syndrome occurs once in approximately 800 to 1.200 births and that, between zero and four years of age, the mortality rate is 52 times higher than for the general population and 37 times higher in the first twenty years of life. This high mortality rate is due to the higher incidence of cardiac alterations, infections of the respiratory system, among other complications arising from its own metabolism.<sup>1,3</sup>

The life expectancy of these special patients varies from 35 to 40 years, but it is estimated that 80% of adult patients survive to 55 years of age or older. The diagnosis of Down's Syndrome is based on a series of signs and symptoms, and its confirmation is established by the chromosomal study. Not all the affected population has the same characteristics, being necessary, in the definitive diagnosis, a cytogenetic investigation to identify the karyotype.  $^{\rm 2,4}$ 

The life expectancy of these special patients varies from 35 to 40 years, but it is estimated that 80% of adult patients survive to 55 years of age or older. The diagnosis of Down's Syndrome is based on a series of signs and symptoms, and its confirmation is established by the chromosomal study. The treatment of Down's Syndrome is strictly aimed at maintaining the patient's quality of life, preventing possible complications. We emphasize the importance of family collaboration, of the multidisciplinary team that deals with patients with the syndrome, as well as the dentist's commitment to the dental aspects of these patients. <sup>1,4</sup>

The team of professionals involved in caring for children with Down syndrome should consider the role of dentistry in achieving better living conditions for this portion of the population. Oral health is still seen as a low priority when compared to medical care dedicated to the individual affected by the syndrome. It is not advisable that professionals who care for patients with Down syndrome leave the problems related to the oral cavity in the background. <sup>3,4</sup>

Considering the various systemic and oral manifestations present in Down syndrome, it is important that the affected population is fully assisted by the health team, including attention to dental aspects. With a view to the practice of comprehensive care, the present study aimed to review articles available in scientific journals that portrayed the dental aspects of patients with Down Syndrome.

#### **METHODS**

The present study is characterized as an integrative review of the literature on

the theme "Down syndrome and dental aspects". Data collection was carried out between the intervals from 2008 to 2018, and the databases used were Brazilian Digital Library of Theses and Dissertations (BDTD), Coordination for the Improvement of Higher Education Personnel (CAPES) and Google Scholar . The strategy of searching for articles in the cited databases was to use the descriptors: "Síndrome de Down, Odontologia, Manifestações bucais".

In the first phase, the articles found underwent the first filtering referring to those available in their entirety, published between 2008 and 2018, in Portuguese and English. In the second phase, the filters independently analyzed whether they were consistent with the theme addressed in the present study, with data such as: author's name, year, objectives, results and discussion. At the end, five articles were selected for analysis in the BDTD, six articles in Google Scholar and four articles in CAPES, totaling, in the end, 15 works used.

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Chart 1: Analysis of the search strategy for articles in the cited databases.						
	TOTAL FOUND	FIRST FILTERING	SECOND FILTERING			
BDTD	143	49	5			
Google Scholar	272	124	6			
CAPES	225	27	4			

Chart 2. Descriptive analysis of scientific productions about Down Syndrome and dental aspects. Caxias- MA, 2018.

TITLE OF THE	AUTHOR/	YEAR OF	ODIECTIVE	TYPE OF	DECUITE
ARTICLE	AUTHORS	PUBLICATION	OBJECTIVE	STUDY	RESULTS
Growth of children and adolescents with Down Syn- drome - A brief literature review	Gorla et al	2011	Perform a literature re- view on growth curves of subjects with Down Syndrome (DS)	Literature review	The results show an approximate growth of -1,5 to -4 standard devia- tions in subjects with DS, when compa- red to the typical population, starting this difference during the prenatal phase and extending to adulthood. There are no major changes between studies of different nationalities.
Special Care Dentistry Association consensus statement on sedation, anesthesia and alternative techniques for people with special needs.	Glassman et al.	2009	Focus on the decision-making process for choosing a treatment method or a combination of methods to facilitate dental treatment for these individuals.	Exploratory descriptive study	Many of these individuals require additional support in addition to local anesthesia to receive dental treatment services. The purpose of this consensus statement is to focus on the decision-making process for choosing a treatment method or a combination of methods to facilitate dental treatment for these individuals.
Knowledge of mo- tor skills and clinical factors of children with Down Syndro- me and the burden of their caregivers.	Lace; Martins	2014	To evaluate the motor performance and clinical characteristics of children with Down syndrome and the mo- thers' knowledge about their children's motor performance.	Exploratory descriptive study	The group of mothers of children with Down syndrome comprised a sample with a mean age of 28 ± 4,5 years, in 55% the pregnancy was not planned and 95% had knowledge about the diagnosis after birth, 80% without complications during pregnancy and 45% have never heard of Down syn- drome. Orthopedic changes were the most prevalent and the burden on the quality of life of mothers in the groups of children with Down syndrome was moderate (22,5 ± 10,6).

Down syndrome: aspects related to the stomatognathic system.	Carvalho, Campos, Crusoé- -Rebello <sup>3</sup>	2010	Conduct a literature review on the main characteristics of the stomatognathic system presented by patients with Down Syndrome, highligh- ting the participation of the dentist in a multidisciplinary team responsible for health promotion.	Revisão de Literatura	The importance of family awareness about the need for oral hygiene for these patients is emphasized, as well as the knowledge of the dental surgeon about the main oral manifestations that affect patients, so that the appropriate treatment is offered and the quality of life of these individuals preserved.
Uso de serviços odontológicos por pacientes com sín- drome de Down	Oliveira et al.,	2008	To analyze the factors related to dental care received by children and adolescents with Down Syndrome.	Estudo de cor- te transversal	Most syndromic patients (79,5%) had already been to the dentist at least once (90% CI: 72,3; 87,8). The children's dental experience was associated with the variables: mothers who claimed to re- ceive guidance from a professional, who assists their child, to take them to the dentist (OR = 6,1 [2,5; 15,1]), children/ adolescents with a previous history of surgery (OR = 2,5 [0,9; 7,1]) and age between 12 and 18 years (OR = 13,1 [2,0; 86,9]).
Importance of an early approach in the dental treat- ment of patients with special needs.	Oliveira; Giro.	2010	Perform a literature review regarding the importance of an early approach in the dental treatment of patients with special needs.	Revisão de Literatura	The dental surgeon must be prepared to care for patients with special needs through technical training and cultivation of human values, minimizing the various problems that these patients are already conditioned to face due to their state of disability.
Evaluation of dental treatment for patients with special needs under general anesthesia.	Castro et al.,	2010	Evaluate the dental tre- atment of patients with special needs under general anesthesia.	Estudo exploratório descritivo	Most patients (98,32%) were classified as ASA II. In the 119 medical records it was found that: in 92 patients (77,31%) 501 restorations were performed; in 89 (74,79%) 602 extractions; in 69 (57,83%) supra/subgingival scraping; in 37 (31,09%) 100 pit and fissure sealants; in 2 (1,68%) 4 pulpotomies; in 1 (0,84%) 1 pulpectomy, and in 1 (0,84%) gingivec- tomy. Review consultations for preventi- ve maintenance were carried out mainly in an outpatient setting (83,10%).
Perspectives for approaching children with Down Syndrome: an integrative litera- ture review from the University of Brasilia.	Santos	2014	To present the scientific evidence available in the literature on nur- sing care for children with Down Syndrome in the period from 2009 to 2013.	Revisão de Literatura	The articles addressed three themes that were easy to perceive: direct attention to the child, attention to the family and the work of health professionals. The attention to the family was the topic that presented the largest number of publi- cations, due to the dependence of every child in relation to their parents being something expected and an even greater dependence on a child with DS, thus, most publications turned to dealing with parents and, through them, enable care.

Dental care pro- gram for special patients: a 13-year experience	Marta	2011	Share the 13-year experience of the Dental Assistance Program for the Special Patient at the Sagrado Coração University, addres- sing the treatment protocol used for both outpatient and hospital treatment, under ge- neral anesthesia with integrated performan- ce of a multidisciplinary team involving under- graduate students in Dentistry	Quantitative study	23,646 surgical-restorative and preventive procedures; families' commitment to the oral health program; marked involvement of undergraduate dentistry students.
Oral health of patients with intel- lectual disabilities	O'Keefe	2010	Review the literature on the oral health of adults with intellectual disabilities.	Literature review	Os estudos eram variáveis qualidade com muitos sendo de um tamanho de amostra pequeno. Pessoas com identidade eram encontradas para ter má higiene bucal e maior prevalência e maior gravidade da doença periodontal. As taxas de cárie em pessoas com ID são de igual ou inferior à população em geral. As taxas de não tra- tado cáries são consistentemente maiores em pessoas com DI
Dental hygiene education about patients with spe- cial needs: a survey of U.S. programs.	Dehaitem et al.,	2008	Explore how dental hygiene programs in the United States educate their students about the treatment of patients with special needs.	Cross-sectio- nal study	Respondents identified "curriculum over- load" as the biggest challenge in meeting patients' special needs. However, 29,4 percent of respondents indicated that they support an increase in clinical experiences for students to give them more opportu- nities to work with patients with special needs.
Cost as a barrier to dental care among people with disabilities: a report from the Florida behavioral risk factor surveillance system.	Rapallo et al.,	2010	Report the cost of care as a barrier to oral health.	Literature review	More people with disabilities reported not seeing a dentist due to the cost than people without disabilities (30% vs. 16%). After ad- justing for confounding variables, disabled Floridians were 60% more likely to report cost as a barrier to dental care (OR = 1,60, 95% Cl 1,32-1,94).
Factors affecting the oral condition of patients with severe motor and intellectual disabi- lities.	Idaiara et al.,	2008	To investigate the specific factors related to the occurrence of dental caries and tooth extractions in patients with severe motor and intellectual disabilities (SMID) residing in an institution.	Cross-sectio- nal study	Through multivariate analysis, rumina- tion and tube feeding were identified as significant factors associated with new tooth decay. On the other hand, childhood or childhood deficiency and salivation were identified as significant factors related to tooth extraction.

Oral health of patients intellec- tual disabilities: a systematic review. Spec Care Dent.	Anders, Davis	2010	Determinar se existem diferenças na saúde bucal entre adultos com deficiência intelec- tual (ID) e a população em geral.	Systematic review	Caries rates in people with ID are equal to or lower than in the general population. However, rates of untreated caries are consistently higher in people with ID. Two subgroups of particularly high risk for oral health problems are people with Down syndrome and people unable to cooperate with routine dental care.
Tooth-brushing intervention programme among children with men- tal handicap	Stefano- vska et al.,	2010	Provide tooth brushing intervention in children with mental disabilities.	Cross-sectio- nal study	For comparative analysis of the OHI database levels and after six months of the intervention program, we found that the average level of the OHI database index for children with mental disabilities is 2,46, and at the end of the program (after six months) was 0,73. The levels of the CPITN index at the beginning and after six months of programmed intervention for children with mental disabilities in both age groups, also confirmed statistical significance for this parameter examined, with an evident reduction in the mean CPITN levels from 2,11 to 0,95. The correlation between OHI levels based on dates and the levels at the end of our in- tervention program means a high positive correlation between these index levels in the initial and final exams.

#### DISCUSSION

Each cell of an ordinary individual has 46 chromosomes, which are divided into 23 pairs, in the individual with Down Syndrome, pair number 21 has one more chromosome, resulting in 47 chromosomes. Gorla et al., (2011)<sup>4</sup> portrays in his study that there are three types of variant anomalies in DS, the most common being trisomy of chromosome 21 due to nonmeiotic disjunction, which occurs in about 95% of patients; the second type is translocation, affecting approximately 3% of carriers; and the third type is known as mosaic, with an incidence of 2%.

According to Glassman and colleagues (2009) <sup>5</sup>, some aspects may be associated with the expansion of the possibility of the birth of a child with DS, among them we can mention the maternal age of 35 or more, because from that age, the risk of having a child with chromosomal abnormality doubles every two years and half, although the birth of children with Down syndrome among young mothers is not ruled out; the paternal age of 45 years or more; previous birth of a child with DS or another chromosomal abnormality; balanced chromosomal translocation in one parent; and parents with chromosomal disorders.<sup>1,2</sup>

To Lace and Martins (2014)<sup>6</sup>, prenatal diagnosis is possibly performed by means of fetal karyotyping. Down syndrome can also be diagnosed after the child's birth due to the recognition of its main phenotypes, such as: generalized muscle hypotonia, flattened occiput, short and thick neck, single fold in the palms of the hands, impairment in length and mental retardation. However, as mentioned in the study by Carvalho, Campos and Crusoé-Rebello (2010)<sup>7</sup>, there is still little knowledge of the pathology by family members who do not always know that the child has this syndrome.

A number of associated pathologies interfere with the development of the-

se individuals. Regarding this information, the study by Oliveira et al.,  $(2008)^8$ shows that patients with down syndrome need exclusive exams to be diagnosed, such as hearing abnormalities (in approximately 80% of cases), orthodontic changes (80 %), vision abnormalities (50%), cardiac abnormalities (40 to 50%), endocrinological changes (15 to 25%), abnormalities of the locomotor system (15%), neurological changes (8%), hematological changes (3%), anomalies of the digestive system (12%), among others. <sup>5</sup>

In the study by Oliveira and Giro  $(2011)^{9}$  it is evident that the knowledge of Down Syndrome is of great interest to the scientific community, as its carriers present a series of craniofacial and dental changes. It is estimated that, in Brazil, only a few dentists are qualified to serve this group of special patients, whose treatment is hampered by little knowledge about their main oral characteristics. <sup>6,8</sup>

As exposed by Castro et al., (2010)<sup>10</sup>, the general characteristics of patients with this syndrome are: flattened faces, Mongoloid orientation of the eyelid clefts, epicanthus, small saddle nose, ear deformities, brachycephaly, flattened occipital region, short and flattened neck, short stature, small and wide hand and feet. Changes in the endocrine-metabolic system, mainly involving the thyroid and pituitary glands, and the hematological and gastrointestinal system, in addition to congenital heart disease and sleep apnea, are also reported. <sup>3,9</sup>

Santos (2014)<sup>11</sup> reports that Down Syndrome is an autosomal congenital condition in which impaired motor coordination, reduced intelligence quotient (IQ), altered facial development due to muscle hypotonia, which affects even the stomatognathic (oral) system, hyperflexibility of the entire body (in infants and early childhood) and premature aging.

To Marta  $(2011)^{12}$ , among the organs that make up the stomatognathic system, the teeth, tongue, periodontium, maxilla, mandible, occlusion, in addition to the temporomandibular joint, may be altered in the patient with chromosome 21 trisomy. The syndromic patient may have macroglossia and a fissured tongue.<sup>7</sup>

Macroglossia, which is characterized by excessive muscle growth and has a congenital origin, can determine tooth displacement and malocclusion. <sup>10</sup> Therefore, Marta (2011) <sup>12</sup> mentions in his study that compensatory mechanisms lead to protrusion and consequent mouth opening, leading the patient to the condition of mouth breather. The surgeries are generally not recommended, because, in addition to the central problem not being the tongue, but the oral cavity, they can interfere with the taste and not help in improving the joint.

The cleft tongue is characterized as a malformation that is manifested clinically by numerous small grooves or grooves on the dorsal surface, which radiate from the central groove of the tongue. Another important factor related to muscle hypotonicity found in the Down Syndrome patient, described in the study by Castro et al. (2010) 10, it is the excess of saliva in the oral commissures, which leads to irritation and fissures, facilitating the installation of infectious processes by the accumulation of microorganisms such as Candida albicans, which is associated with the lack of local hygiene.<sup>12</sup>

> The teeth of Down Syndrome patients show complete mineralization, but, in addition to being delayed, they show alterations in the eruption sequence, mainly of the deciduous ones.

Certain population groups are more susceptible to periodontal disease, such as special patients, including Down Syndrome patients. O'keefe (2010)<sup>13</sup> states in herwork that the direct effects are the microbial factors that act directly on the tissues causing damage to them, and the indirect effects are the self-injurious factors produced by the host, such as inflammatory responses and immunological phenomena, in response to bacterial aggression.<sup>14</sup>

Regarding craniofacial aspects, micrognathia is observed, characterized by a small jaw, which may occasionally affect the mandible. The micrognathia of the jaw, according to O'keefe (2010)<sup>13</sup>, is usually due to a deficiency in the premaxillary area, and patients with this deformity have the middle third of the face retracted. It is believed that micrognathia is the main responsible for the development of mouth breathing, the main mouth characteristic, due to the association with changes in the development of nasal and nasopharyngeal structures. <sup>10</sup>

Regarding the mandible, as depicted by Dehaitem et al (2008) <sup>14</sup>, it can be expanded transversely due to lingual pressures. As a consequence, the patient has temporomandibular joint disorders associated with generalized muscle hypotonicity and alteration of the articular disc. The chronology of eruption of primary and permanent teeth is varied, often delayed, and usually the primary dentition is not complete until the age of four or five. <sup>5,13</sup>

The teeth of Down Syndrome patients show complete mineralization, but, in addition to being delayed, they show alterations in the eruption sequence, mainly of the deciduous ones. Rapalo et al (2010) <sup>15</sup> highlights that microdontia and enamel hypoplasia, as well as hypodontia and oligodontia, are the most common dental anomalies. Structural anomalies include taurodontism, conoid teeth, fusions and germinations, the canines being most affected in shape and size. <sup>1,4</sup>

It is also observed in patients with Down syndrome, warhead palate, hypertrophied tonsils and adenoids, bifid uvula, cleft lip and palate. Malocclusion is frequently found in patients with Down syndrome, with Angle class III predominating, posterior crossbite, pseudo prognathism and anterior open bite. According to the study by Idaira et al.,  $(2010)^{16}$ , the increase in the individual's age is related to the tendency to develop malocclusion and the combination with other problems such as oral motor dysfunction, reduced muscle tone in the mouth and face, which can lead to speech, chewing and swallowing changes. <sup>15</sup>

To Anders and Davis (2010)<sup>17</sup>, bruxism is also observed, being more common at night, leading to the consequent uniform wear of tooth enamel. The position of the teeth also plays an important role in certain circumstances, favoring the accumulation of food and waste. If we consider that, in general, patients with special needs have poor oral health and poor hygiene, we could conclude that the syndromic patient has a high rate of caries. However, this does not happen, since macroglossia and the salivary process are factors that determine the low rate of caries.

Finally, Stefanovska et al., (2010)<sup>18</sup> states that in addition to immunological changes, other factors are proposed to explain the high prevalence of periodontal disease and the increase in its severity in patients with the syndrome, including malocclusion, early colonization and a high number of periodontopathogens, especially Porphyromonas gingivalis.<sup>21,6</sup>

#### CONCLUSION

From the information obtained, it is concluded that Down syndrome is a multisystemic congenital disease, associated with several craniofacial and dental abnormalities, mental retardation and physical malformations. Patients with trisomy 21 may be treated clinically by dental professionals, provided the necessary care is taken. Supervised oral hygiene is essential in these individuals, due to motor limitations and the high prevalence of periodontal diseases. The correct assessment of growth and development, especially in the first years, allows the detection of problems that can impede the full development of the individual. Regular follow-up by dentists, in order to promote the necessary well-being in the various areas of human performance, aims at enriching the patient's quality of life.

It is exposed that it is necessary to encourage the publication of more scientific articles in the area. It is opportune to consider the works already described in the academic community, in order to strengthen future research on this topic. As the article reveals, there are several authors who try to make clear the importance of the role of the dental professional in monitoring people with Down Syndrome.

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