

Frequent oral manifestations in people with Down Syndrome. A literature review.

Manifestaciones orales frecuentes en personas con Síndrome de Down. Una revisión de la literatura.

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INTRODUCTION.

Down syndrome (DS) is one of the most common genetic disorders in the world, with an estimated incidence of 1 per 600-1,000 live births.¹ Its origin is in 95% of the cases due to the presence of an additional copy of chromosome 21, followed by translocation (2%–4%) and isochromosomal mosaicism (1–2%).²

The cognitive functions of individuals with DS range from mild to moderate intelligence quotient (IQ). In addition, a certain incidence of delay in expressive language is reported, which poses challenges to access to health care services.³

The literature indicates that this condition predisposes to certain alterations such as an abnormal position of the tongue, craniofacial deformities, dental alterations and muscular disorders,⁴ in addition to a high prevalence of gingivitis and periodontal disease,⁵ among others. Therefore, the purpose of this literature review is to describe the most frequent oral manifestations in people with Down Syndrome.

MATERIALS AND METHODS.

A literature review was carried out from articles collected from three databases PubMed, Web of Science and Scopus using the following terms: Down's syndrome, trisomy,²¹ dental, oral, decay, demineralization, periodontal, periodontitis, gingivitis, soft tissue, lips, tongue, macroglossia, occlusion, hypodontia, class III, palate, ASD, autism.

Complete original articles were selected from the last 10 years written in English, and carried out in humans. All the included studies were observational studies, systematic reviews and meta-analyses, which had information related to oral pathologies or conditions and whose central study population were individuals with DS.

Narrative review articles and case reports were excluded from the search, as well as those describing the perception of caregivers regarding the oral health status of individuals with DS and articles whose objective was to quantify bacterial species, cytokines, salivary

components, among others and were not primarily focused on oral pathologies or conditions. Articles whose full text could not be obtained were also excluded.

RESULTS.

Fifteen articles were obtained that met the inclusion and exclusion criteria.

The countries of origin of the studies included Brazil,^{2,4,6-8} Yemen,^{9,10} Saudi Arabia,¹¹ United Arab Emirates,³ India,¹² Indonesia,¹³ Jordan,¹⁴ Kosovo,⁵ the Netherlands,¹⁵ and Peru.¹

Six articles were published between the years 2011-2015 and nine between 2016-2020. There are two systematic reviews that are meta-analyses and 13 articles that are observational studies.

Table 1. Randomized clinical trials included in the review.

Authors (years)	Study N	Age (year)	Characteristics	Results (%)
Al Habashneh <i>et al.</i> ¹⁴ (2012)	DS: 103 C: 103	12-16	Malocclusion Fissured Tongue* Caries Hypodontia* G. spacing* Clase III Crowding Open Bite Oclusal wear Peg laterals Hypoplasia * Microdontia Fusión*	DS: 69.9 / C: 40.8 DS: 56.3 / C: 2.9 DS: 56.3 / C: 49.8 DS: 51.5 / C: 4.9 DS: 51.5 / 15.5 DS: 47.5 / C: 11.65 DS: 37.9 / C: 31.1 DS: 35.9 / C: 4.9 DS: 35.0 / C: 11.7 DS: 20.4 / C: 2.9 DS: 8.73 / C: 1.94 DS: 0.9 / C: 0 DS: 0.9 / C: 0
Al-Sufyani <i>et al.</i> ⁹ (2014)	1101	6-16	Gingivitis	100 (28.7 severe; 47.5 moderate; 23.8 mild)
Shukla <i>et al.</i> ¹² (2014)	77	6-40	Malocclusion High arched palate Caries Fissured Tongue Macroglossia Lack of lip seal Microdontia Clase III Crossbite Congenitally missing teeth Crowding of anterior teeth Angular Cheilitis Open Bite Ankyloglossia	97 84.4 78.0 67.5 58.4 51.9 45.5 42.9 33.8 33.8 23.4 22.1 19.5 13.0
Al-Maweri <i>et al.</i> ¹⁰ (2015)	C:50 C:50	6-18	Gingivitis Fissured Tongue* Lip fissures* Angular Cheilitis * Gingival hyperplasia * Cheilitis * Fibroma Traumatic ulcer Herpes labialis	DS: 96 / C: 90 DS: 78 / C: 24 DS: 64 / C: 0 DS: 38 / C: 2 DS: 18 / C: 0 DS: 14 / C: 2 DS: 8 / C: 0 DS: 4 / C: 2 DS: 2 / C: 2

*: Statistically significant difference. DS: Down Syndrome. C: Control. DMFT: Decayed, Missing, and Filled Teeth index. [Table 1 continued].

Marques <i>et al.</i> ⁴ (2015)	DS:60	S:x 14.7	Missing teeth (at least one)	DS: 95.5 / C: 4.5
	C:60	C:x 12.18	Clase III*	DS: 88.6 / C: 11.4
			Anterior Open Bite (≥ 2)*	DS: 84.2 / C: 15.8
			Posterior Crossbite*	DS: 82.1 / C: 17.9
			Spacing (1 or 2 segments)	DS: 48 / C: 52
			Crowding (1 or 2 segments)	DS: 43.1 / C: 56.9
Van Marrewijk <i>et al.</i> ¹⁵ (2016)	63	6-19	Hypodontia	60.3
			Oligodontia	23.8
Aparecido <i>et al.</i> ⁷ (2016)	105	7 - 42	Hypodontia	16.19
			Microdontia	16.19
			Retained Teeth	10.47
			Taurodontism	9.52
			Supernumerary Teeth	5.71
			Macrodontia	2.85
			Root dilaceration	0.95
Begzati <i>et al.</i> ⁵ (2017)	65	6-18	Gingivitis	65
			Third molar agenesis	63
			Hypodontia	52
			Mandibular prognathism	48
			Periodontitis	43
			Anterior crossbite	37
			Anterior openbite	25
			Posterior crossbite	15
			Teeth transposition	12
			Supernumerary teeth	5
Anggraini <i>et al.</i> ¹³ (2019)	174	14-53	Hypodontia	47.7
			Microdontia	47.7
			Enamel hypoplasia	10.3
			Supernumerary Teeth	9.8
			Enamel hypocalcification	2.3
			Fusion	2.3
			Talon cusp	1.1
			Macrodontia	0.6
Ghaith <i>et al.</i> ³ (2019)	DS:106 C: 125	4-18	High arched palate	DS: 93.4 / C: 33.6
			Deep bite	DS: 90.6 / C: 18.4
			Scissor bite	DS: 90.5 / C: 2.4
			Posterior Spacing	DS: 79.2 / C: 8.0
			Fissured Tongue*	DS: 67.9 / C: 0.0
			Clase III*	DS: 66.0 / C: 11.2
			Gingivitis	DS: 65.4 / C: 70.4
			Open bite	DS: 60.0 / C: 11.2
			Anterior Spacing	DS: 54.7 / C: 32.0
			Cross bite	DS: 50.9 / C: 28.0
			Macroglossia*	DS: 46.2 / C: 0.8
			Microdontia	DS: 45.3 / C: 0.8
			Dental erosion *	DS: 34.0 / C: 15.3
			Angular cheilitis*	DS: 21.7 / C: 0.0
			Geographic tongue*	DS: 8.5 / C: 0.0
			Atrophy of the tongue*	DS: 7.5 / C: 0.0
			Ulcers	DS: 2.8 / C: 0.0
			Trauma of soft tissue	DS: 2.8 / C: 0.0
			Irritation fibroma	DS: 1.0 / C: 0.0
			Mean dmft	DS: 3.42 / C: 2.76
			Mean DMFT*	DS: 3.32 / C: 2.16

*: Statistically significant difference. DS: Down Syndrome. C: Control. DMFT: Decayed, Missing, and Filled Teeth index. [Table 1 continued].

Nuernberg <i>et al.</i> ² (2019)	64	14-51	Gingivitis Periodontitis	28.1 71.9 (62.5 localized; 9.4 generalized)
Tipe <i>et al.</i> ¹ (2019)	107	12-16	Periodontitis Clase III Gingivitis	85.9 mild – 2.8 moderate 69 58.8 mild – 29.8 moderate
Assery <i>et al.</i> ¹¹ (2020)	DS: 30 C: 30	7-12	Clase III* Open Bite* Primary dentition caries Permanent dentition caries *	DS: 36.7 / C: 0 DS: 40 / C: 3.3 DS: 66.7 / C: 56.7 DS: 20 / C: 56.7

*: Statistically significant difference. **DS:** Down Syndrome. **C:** Control. **DMFT:** Decayed, Missing, and Filled Teeth index. [Table 1].

The information obtained from the 13 observational articles is summarized in Table 1.

DISCUSSION.

The increase in life expectancy of individuals with DS¹⁵ in recent decades makes it important to identify the frequent oral manifestations of this population. The reviewed studies indicated that malocclusions are highly prevalent in people with DS, class III malocclusion being the most frequent.^{1,3-5,11,12,14}

A high prevalence of open bite^{3-5,11,12,14} and cross bite^{3-5,12} are also reported. This is in agreement with Doriguetto *et al.*,⁸ who carried out a systematic review with a meta-analysis indicating that, when compared to controls, children and adolescents with DS had a greater presence of class III, posterior/anterior cross bite and anterior open bite. The high risk of malocclusion may be related both to the genetic predisposition of people with DS and to the interaction between muscle function and skeletal development during growth.¹¹

Macroglossia, fissured tongue and angular cheilitis were the most frequent soft tissue alterations in individuals with DS,^{10,12,14} There was no concordance in other soft tissue conditions.

Regarding dental anomalies, one of the most reported was hypodontia^{5,7,13-15} followed by microdontia^{3,7,12-14} and shape anomalies.^{7,13} Literature considers that dental anomalies in individuals with DS should not be considered as an isolated phenomena,

but as part of a set of genetically determined characteristics, possibly associated with the slow rate of cell growth and the consequent reduction in the number of cells.⁷ Gaith *et al.*,³ and Al-Maweri *et al.*,¹⁰ compared the gingival status of people with DS and controls, finding that the presence of gingivitis was similar in both groups.

Among the reviewed articles that studied periodontal tissues but did not have controls, there were a high prevalence of gingivitis and periodontitis.

Literature describes increased susceptibility to periodontal disease observed in subjects with DS associated with various factors, including early microbial colonization, microbiota composition, impaired immune response, and increased gingival inflammation.² When comparing caries prevalence in DS compared with controls, the results of the included articles were contradictory.

Habashneh *et al.*,¹⁴ found no significant differences between both groups. Gaith *et al.*,³ and Assery *et al.*,¹¹ also found no significant differences for primary dentition. However, for permanent dentition Gaith *et al.*,³ found a higher caries damage in DS, contrary to Assery *et al.*,¹¹ who described a higher prevalence of caries in control groups.

A systematic review with meta-analysis⁶ found limited scientific evidence suggesting that people with DS have less caries than people without DS, but that this evidence may be weakened by a lack of control for confounding factors.

The multifactorial nature of dental cavities may explain the diversity of the results.

CONCLUSION.

The results of this literature review show a high prevalence of class III malocclusions, open bite, cross bite, macroglossia, fissured tongue, angular cheilitis, agenesis/hypodontia, dental anomalies, gingivitis and periodontitis in the population with DS. The large number of oral manifestations associated with DS makes it necessary for dentists to have up-to-date knowledge on the subject in order to carry out treatments with a preventive approach.

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