

## THE IMPORTANCE OF PREVENTIVE ORTHODONTICS IN DOWN SYNDROME.

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Os autores alegam não haver conflito de interesses.

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### Abstract

**Introduction:** Down Syndrome (DS) is an autosomal genetic condition of the chromosomal pair 21 that shows craniofacial characteristics, cognitive deficit, and generalized muscular hypotonia. These characteristics have implications for eating, swallowing, chewing, posture, breathing, and sleep. Early orthodontic intervention can improve orofacial functions by strengthening and stimulating facial muscles. The objective of the study was to make a literature review on the general and oral aspects of patients with Down's Syndrome, showing possible treatments for their orofacial rehabilitation, highlighting the importance of early orthodontic treatment

for these individuals, as well as the benefits of these interventions in the quality of life. Data source: A survey of articles published between years 2011 to 2017 was carried out by means of search strategy based on terms in Portuguese: "Síndrome de Down"; "Ortodontia"; "Má Oclusão," "Qualidade de vida" and in English: "Down Syndrome", "Orthodontics"; "Malocclusion", "Quality of Life". Conclusion: The orthodontic intervention performed with several devices in a preventive way will help children with DS in a better bio-psycho-social development, with a positive influence on the quality of life of these individuals.

**Key words:** Down Syndrome; Orthodontics; Malocclusion; Quality of Life.

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## INTRODUCTION

Down Syndrome (DS) is an autosomal genetic condition of chromosome 21 pair and is considered a very frequent alteration, affecting 1 in 700 live births.<sup>1</sup>

They have specific characteristics such as cognitive impairment, generalized muscular hypotonia, mid-third underdevelopment, brachycephaly, ear pinna malformation, oblique eyelid clefts, epicanthus, flattened nasal base, short limbs, and single transverse palmar fold.<sup>1,2</sup>

Patients with DS may present with dental alterations such as hypodontia, conoid or microdontic teeth, enamel hypoplasia, fusion, twinning, taurodontia, delay or reversal of the eruption order of the deciduous teeth<sup>2</sup>. The palate presents ogival shape and reduced dimensions, while the tongue has relative macroglossia.<sup>3,4</sup>

Skeletal patterns II and III in which underdevelopment of the middle third of the face that provide mandibular projection are among the craniofacial characteristics frequently found in DS. The most commonly found malocclusions are the class III and II molar relationships, anterior and posterior crossbites, and anterior open bite.<sup>4,5,6</sup>

Changes in the masticatory system of patients with DS have implications for eating, swallowing, chewing, phonation, posture, breathing, sleep, and social impacts, as well as problems with self-esteem and aesthetics of children and adults.<sup>5,6</sup>

Orthodontics in Down Syndrome aims to improve orofacial functions by strengthening and stimulating the muscles of the face (Castillo Morales Palatal Memory Plaque, Trainer System). Craniofacial changes can be corrected with transverse maxillary enlargement, positioning of primary and permanent teeth with braces or mobile appliances.<sup>7,8,9,10,11</sup>

The aim of this study is to perform a literature review regarding the general and oral aspects of Down Syndrome patients, exploring possible treatments options for their orofacial rehabilitation, highlighting the importance of early orthodontic treatment, and the benefits of these interventions for

improving quality of life.

Articles published between the years 2010 and 2017 were surveyed through search strategy based on the following terms: Down Syndrome, Orthodontics, Malocclusion, Quality of Life. The abstracts of the retrieved articles were analyzed to verify compliance with the inclusion and exclusion criteria. The following inclusion criteria were adopted: articles published in Portuguese and English and indexed; articles that contemplated malocclusions and their ways of treatment. Exclusion criteria were: studies without information on the sampling and analysis performed; theses, dissertations and articles that addresses other types of malocclusion rather than the one studied.

## LITERATURE REVIEW

Patients with disabilities or special needs may present a great challenge in the dental routine<sup>8</sup>. "Your medical complexity associated with medications you usually use can affect your oral health". These individuals require differentiated accommodation and management to treat their oral health.<sup>12,13,14</sup>

Trisomy 21, or Down Syndrome, is the most common chromosomal disorder in mankind with a prevalence of one case in 700 live births<sup>2,12</sup>. These children can present with congenital heart disease, gastrointestinal disorders, visual and hearing impairment, generalized muscle hypotonia, cognitive impairment, and growth disorders.<sup>15,16</sup>

Individuals with DS show a high prevalence of obstructive sleep apnea due to underdevelopment of the middle third of the face, narrow nasopharynx, tonsil and adenoid hypertrophy.<sup>12,17</sup> The tongue is fissured and presents hypotonia; therefore, its movement is slow and inaccurate. Lip sealing is also insufficient due to decreased lip tonicity.<sup>6,17</sup>

The most common dental alterations found in DS are delayed eruption of primary teeth, number (agenesis), shape (fusion, gemination), size (microdontia and taurodontia), and structure (hypocalcification of enamel). The palate has an ogival and deep shape, with higher prevalence of malocclusion (Figure 1).<sup>18,19</sup>



Figure 1: DS patient, 3 years old male, with lower incisor microdontia, various diastemas, and right and anterior crossbites.

With the underdevelopment of the middle third of the face, the oral and nasal cavity are small and many children with DS are oral breathers.<sup>20</sup> The flaccid and hypotonic tongue does not find enough space to remain well positioned (i.e., resting on the palate behind the upper incisors), settles low on the mandible, and, in many patients, rests between the lower lip and teeth. This poor posture of the oral muscles and tongue is one of the beginning and perpetuating factors of malocclusions in individuals with Down Syndrome.<sup>15,20,21</sup>

The maintenance of functional and anatomical integrity of the Stomatognathic System allows the correct growth and development of intra and extraoral structures. Stimulation of speech and craniofacial development begins in the first year of life of DS children, and include Speech Therapy and consultation with an Orthodontist.<sup>8,22</sup>

When evaluated early by a multidisciplinary team, infants and children with DS may have the indication of a palatal plate<sup>16,17</sup>. This device is used preventively during the first months of life to aid in the neuromotor rehabilitation of the orofacial

region. Rodolfo Castillo Morales, Argentine, developed the palatal memory plate (PPM) for children diagnosed with predominantly oral breathing, muscle hypotonia, lingual protrusion, and insufficient lip sealing.<sup>9,17</sup>

The plates used vary according to age of the children and their characteristics. They are made out of self-curing acrylic molded by the dentist, and must have a tongue stimulation zone (openings or beads) and an upper lip activation area (strips, or buttons).<sup>22, 23</sup>

The palatal plate creates sensory and motor stimulation of the tongue and lips. The projections and openings create a reflex in the tongue musculature by stimulating it to contract and move up and posteriorly in the mouth, promoting contact with the palate, and thus increasing tongue activity (Figure 2).<sup>23</sup>

The installation of the intraoral device, together with speech therapy, promotes stimulation of specific tongue movements, increase on upper lip mobility and facial muscle tone, lip sealing, improvement in breastfeeding, sucking, and swallowing, speech development, and facilitates the development of nasal breathing.<sup>8,9,12</sup>





Figure 2: Palatal plate for children with teeth, with expander vise to assist in the correction of anteroposterior underdevelopment. The space created in the plate serves to stimulate the tongue.

The malocclusions found in DS patients are complex due to the involvement of skeletal, dental structures or a combination of both, so that early intervention is of utmost importance.<sup>5,6,7</sup>

Orthodontic intervention in deciduous and mixed dentitions, such as maxillary expansion with or without extraoral traction (face mask) therapy, is believed to reduce bone discrepancies and favor more satisfactory craniofacial and dental

development.<sup>8,11,19</sup>

Maxillary expansion appliances are an efficient orthopedic therapeutic option used for transverse augmentation of the maxillary bone, leading to widening of the arch perimeter. Expansion appliances provide more space for alignment of both deciduous and future permanent teeth, and also promote the expansion of the complex nasomaxillary, resulting in improved nasal ventilation and tongue accommodation (Figure 3).<sup>11, 17</sup>

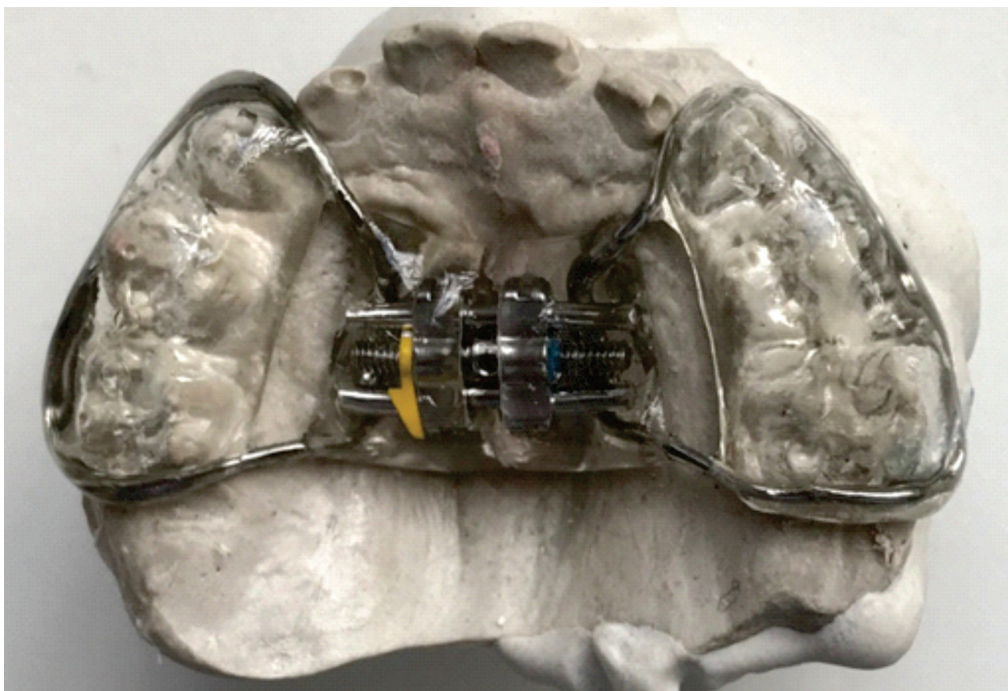


Figure 3: Haas teeth-encapsulated maxilla expander

For maxillary expansion to occur, the force applied to the teeth and the alveolar process has to exceed the limit necessary for orthodontic tooth movement, promoting the separation of the suture from the maxillary bones.<sup>12,19</sup>

Device activation in DS patients differs from those without DS.<sup>14</sup> While activation of typical patients is daily, ranging from 0.3 to 0.5mm, activation of children with Down Syndrome will depend on family routine, patient individuality, associated morbidities such as sensory processing, autism,

deleterious habits, and disorders of anxiety.<sup>8</sup>

The orthodontist determines the amount of activations and estimated treatment time for each patient.<sup>8</sup> The family should be advised that the appliance has to be worn for as long as necessary to deconstruct the bite, allowing for non-painful separation at the midpalatal suture level upper jaw in growing children. After obtaining the desired transverse relationship, the device itself becomes a containment, allowing bone remodeling and consolidation in the suture region (Figure 4).<sup>11,12,19</sup>

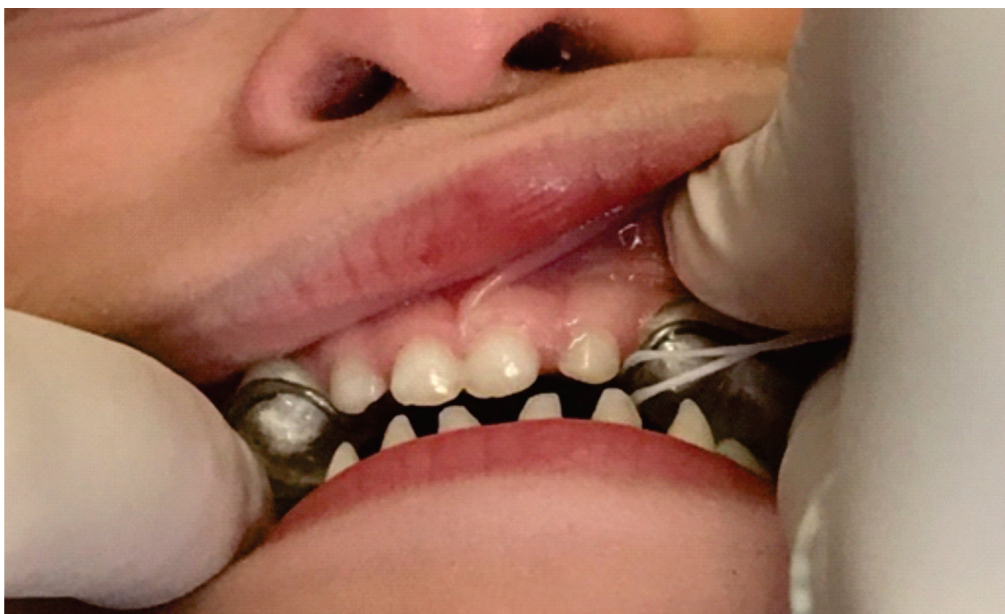


Figure 4: Patient placing Haas expander.

Researcher Dr. Chris Farrell has created the Trainer devices, which represent another therapeutic option for DS patients. The devices (T4KTM - 1st phase and T4KTM - 2nd phase) have the main function of stimulating orofacial functions and thus correcting soft tissue dysfunction, resulting from bad myofunctional habits. This correction leads to improvement in dental occlusion and mandibular posture.<sup>18, 22</sup>

The Trainer system consists of prefabricated polyurethane devices made out of various elements that stimulate the facial, masticatory, and lingual muscles.<sup>22</sup> There are devices indicated for facial pattern II and III patients. The devices for pattern II move the mandible to an anterior position and stimulates the transverse development. The devices for pattern III are indicated for

patients with a tendency to midface atresia and mandibular projection.<sup>22</sup> The Trainer system promotes improved lip sealing, anterior open bite closure, more harmonious facial appearance, and improved tongue positioning in the oral cavity.<sup>6, 22</sup>

## DISCUSSION

Individuals with DS have particularly more dentoskeletal changes than those of the general population<sup>1,4,20</sup>. This is due to the increased frequency of craniofacial alterations, underdevelopment and growth of the maxillary bones, and generalized hypotonia affecting the tongue and orofacial muscles<sup>1,3,5,21</sup>.

Orthodontic and orthopedic preventive procedures may favorably interfere with the

dental treatment, so that patients with DS may not acquire future dental problems<sup>7,8,12,19</sup>.

Some authors<sup>9,16,17</sup> have obtained excellent results with the installation of the Castillo Morales palatal memory plate in children with DS and reported lingual retropositioning, with strengthening of the orbicularis musculature of the mouth, lip sealing, and more satisfactory speech development.

According to the literature<sup>16,17</sup> the use of palatal memory plates (PPM) is well known in neonates with DS, but can also be used in babies diagnosed with Piérre-Robin Syndrome, cleft palate/lip, Moebius Syndrome, and Beckwith-Wiedmann Syndrome.

The time of PPM use is still controversial in the literature. Some authors indicate 5 to 10 minutes, twice a day, while other studies report that the plate should be worn for at least 2 hours<sup>8,12,10,16,17</sup>.

Rapid maxillary expansion (RME) has been unanimously shown<sup>7,8,11,12,14,15,19</sup> to be effective in treating children with constriction and maxillary underdevelopment, and for obstructive sleep apnea.

There are reports in the literature indicating that many patients with maxillary deficiency have a history of respiratory infections since childhood and, thus, are affected by conductive hearing loss<sup>11</sup>. Researchers have indicated that RME, due to rapid results in patients with skeletal changes, can be considered an acceptable treatment to prevent recurrent otitis media in children affected by anatomical maxillary changes. Maxillary expansion extends palate muscles and elevator muscles, helping with restoration of auditory tube function<sup>11,21</sup>.

The most commonly used devices for maxillary expansion are Mcnamara, Haas, and Hyrax<sup>8</sup>. Regarding a protocol after the installation of maxillary expanders in patients with DS, no specific protocol has been found in the literature regarding the amount and time of activation of the device<sup>8,14,15,19</sup>.

The Trainer System, a mobile device technique, can become a challenge when indicated for non-collaborative DS patients, because the treatment outcome depends on the correct use of the device<sup>8,12,14</sup>. Patient

and parents education, motivation, and commitment are keys factors in obtaining successful results.<sup>7,18,22</sup>

The literature is emphatic in saying that children with Down Syndrome should be assisted by a team of speech therapists, occupational therapists, pediatricians, physiotherapists, and dentists<sup>2,20,21</sup>. Orthodontic therapy should be known to health professionals who assist infants and children with DS, so that interventions are indicated early, ensuring benefits of these treatments on the quality of life of those patients.<sup>9,15,20,22</sup>

## CONCLUSION

There are several options for early and effective orthodontic correction of transverse maxillary deficiency and class III malocclusion of Down Syndrome patients.

It is essential that these individuals are assisted and treated by a multidisciplinary team, including orthodontists, physical therapists, speech therapists, occupational therapists, and physicians.

Orthodontic treatment is essential during the life of patients with Down Syndrome, assisting in functional problems such as swallowing, chewing, and phonation, as well as aesthetics, positively influencing the physical, psychological, and social development of those children.

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