

## Non-syndromic oligodontia: A case report

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### Abstract

Oligodontia is the congenital absence of six or more permanent teeth, excluding the third molars. Some terms are used to define the absence of primary or permanent teeth, such as hypodontia or dental agenesis. This condition can occur as an isolated dental feature (non-syndromic) or concomitant with a general disorder (syndromic). The etiology is attributed to genetic (mutations in genes such as PAX9, AXIN2, EDA, MSX1) and environmental factors. The sequelae of oligodontia include missing teeth, prolonged retention of primary teeth, root resorption and ankylosis resulting in unstable occlusion. The space for rehabilitation depends on the pattern and severity of tooth absence.

We present a female (8 years old) with no relevant medical history, with absence of second permanent molars, lower central incisors, class II division 1 malocclusion and delayed emergence of first permanent molars. Management of malocclusion and conservation of spaces was carried out after 6 months of observation.

Early diagnosis by clinical and radiographic examination is essential to assess the number, position of present and absent teeth and the condition of the alveolar ridge during subsequent treatment planning with the multidisciplinary team.

**Keywords:** Oligodontia; Hypodontia; Agenesis; Non-syndromic; Children

### 1. Introduction

The absence of teeth is the most common developmental anomaly in humans. According to the Index of Classification of Diseases for epidemiological purposes, dental agenesis is recognized as the lack of development of a dental germ, which can be classified as hypodontia (absence of one to five teeth), oligodontia (absence of six or more teeth, excluding the third molars), partial anodontia (absence of all of one or both tooth formulas of primary or permanent teeth) and total anodontia (absence of all teeth). According to the authors, the congenital absence of certain teeth could be framed in the process of human evolution [1].

Syndromic oligodontia can occur as a dental trait attributed to mutations in genes: PAX9, MSX1, AXIN2, EDA and EDAR or concomitantly with systemic diseases such as ectodermal dysplasia, Down syndrome, Nance-Horan syndrome, Rieger syndrome, Seckel syndrome, Wolf-Hirschhorn syndrome, Klippel-Feil syndrome, Van der Woude syndrome and cleft lip and palate, among others [2,3,4]. Non-syndromic oligodontia could be due to environmental causes, medications, and nutritional imbalances as isolated factors.

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The prevalence of this entity is 0.09% in the general population [5] but varies according to different geographic regions and ethnicities. In Caucasian populations in North America, Australia, and Europe it is reported to be 0.14 to 7%, 7 and 0.25% in Asian populations in China [6].

The pattern of missing teeth reported in the literature is variable, can be symmetrical or incidental and is frequently associated with other dental anomalies, such as alterations in the size and/or shape of the teeth, alterations in dental maturation, dental development, both by formation and eruption, crowding, malocclusion and altered craniofacial growth [5].

Prolonged retention of primary teeth, infraocclusion, ankylosis, unstable occlusion, microdontia or conical teeth are reported as consequences of the absence of teeth [3]. In addition, the absence of permanent tooth germs can cause atrophy of the alveolar ridge, which can pose problems in subsequent prosthodontic treatment with implants. The rehabilitation depends on the pattern and severity of the missing teeth [3].

The relationship between dental agenesis and the presence of dental or skeletal malocclusions has been described by various authors. The results coincide that depending on the ethnic group, the upper incisors can present pro- or retro inclination, the maxillae can have a reduced size and present a reduction of the vertical dimension being unfavorable both aesthetically and functionally [2][7].

Timely diagnosis of this condition allows for comprehensive treatment planning that includes all phases of dental development, with the planning of short-, medium- and long-term objectives [8].

Treatment should be oriented towards promoting the correct development of occlusion, restoring function and esthetics in patients with this condition. However, treatment planning presents a challenge, since any tooth can be affected and this added to malocclusion; therefore, a multidisciplinary management is performed in the different stages of dental replacement in which the pediatric dentist, the orthodontist and the oral rehabilitator participate; with the objective of maintaining, regaining, and closing spaces for subsequent management once growth is completed [8].

In the deciduous or early mixed dentition, prevention (dietary counseling, fluorides, pit and fissure sealants and mouth guards), removable prostheses for psychological and functional reasons (regular adjustments during growth, retention and stability due to underdeveloped alveolar ridges), composite resin restorations to improve the esthetics of microdontic permanent teeth, interceptive orthodontic treatments to help close spaces (long-term retention may be necessary) and interceptive orthopedic treatment (use of orthopedic appliances for malocclusions) [8][3].

Therefore, the purpose of this report is to show preliminarily the management of a clinical case of non-syndromic oligodontia.

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## 2. Case report

Female patient of 8 years, who came to the office by her own means, with no relevant medical history. Intraoral examination showed a convex profile. Intraoral examination showed delayed eruption of the first permanent molars, right straight terminal plane and left distal step, bilateral posterior crossbite, anterior overbite, overjet 5mm, overbite 90%, vestibularization of upper central incisors (Figure 1).



**Figure 1** Front, profile, upper and lower occlusal photographs. Note the delayed emergence of the first permanent molars, the persistence of the deciduous lower central incisor and the malocclusion

Radiographic examination shows absence of the germs of the upper and lower permanent second molars and lower central incisors (Figure 2).

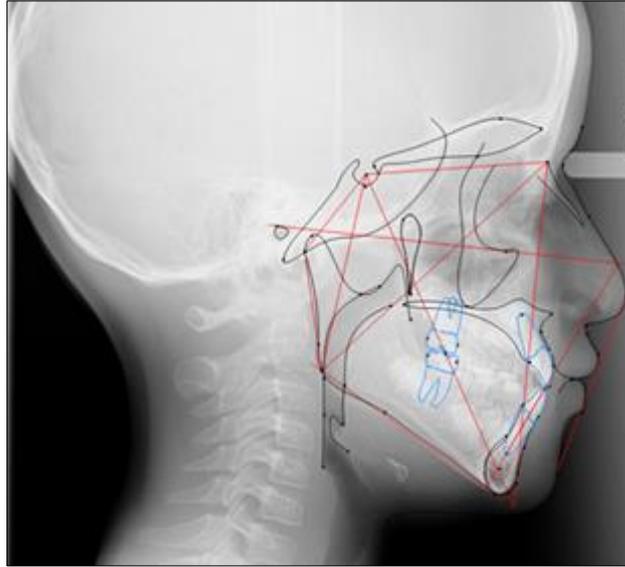


**Figure 2** Panoramic radiograph. Note the absence of permanent teeth

Cephalometric examination showed Class II division 1, mandibular retrognathism, maxillary normognathism, pro inclination of upper incisors, horizontal growth pattern (Figure 3).

The treatment was planned in different phases: preventive, biofilm control, instruction and motivation in oral hygiene, topical application of fluoride, corrective, use of a Class II Twin Block orthopedic appliance (Figure 4) to stimulate the growth of the mandible and maxillary expansion screw due to the malocclusion and conservation of space by maintaining tooth 7.1, and as a maintenance phase, appliance controls every month. The main objective of the treatment is based on improving the patient's quality of life, which allows correcting the malocclusion to improve the facial profile, maintaining the space due to the absence of lower central incisors for subsequent oral rehabilitation after completing the patient's growth.

Monthly check-ups were carried out and at 6 months there was evidence of a better facial profile, mandibular advancement, maintenance of tooth 7.1 (Figure 5) and emergence of the first permanent molars 1.6, 2.6 and 3.6. The patient will continue with subsequent periodic check-ups.



**Figure 3** Cephalometric study



**Figure 4** Class II Twin Block orthopedic appliance



**Figure 5** Extraoral photographs after six months of treatment, note the facial profile

### 3. Discussion

Oligodontia is the congenital absence of six or more permanent teeth, excluding the third molars, and comprises the most severe forms of dental agenesis [9]. Some terms are used to define the absence of primary or permanent teeth, such as hypodontia or agenesis [6].

This condition can occur as an isolated dental trait (non-syndromic) or concomitant with a general disorder (syndromic) [3][10]. Gill et al. in 2015 [8], indicate that oligodontia can be inherited by autosomal dominant, autosomal recessive or sex-linked traits, can have varying degrees of expression and that the higher prevalence in families would suggest a genetic link, unlike the case presented where no family history of dental anomalies of number is referred. The etiology is attributed to genetic factors (mutations in genes such as: PAX9, AXIN2, EDA, MSX1) and environmental factors such as: nutritional imbalances, trauma, infections such as rubella, exposure to drugs such as thalidomide, hormonal influences, previous irradiation, or presence of tumors, among others [4]. Zhou et al. in 2021[6], identified a new mutation in WNT10A (c.99 105dup) and eight mutations previously found in WNT10A (c.433 G > A; c.682 T > A; c.318 C > G; c.511.C > T; c.321 C > A), EDAR (c. 581 C > T) and LRP6 (c. 1003 C > T, c. 2747 G > T) and correlated genotype-phenotype of molar agenesis likely related to PAX9 mutations, lower incisor and upper lateral incisor agenesis more closely related to EDA mutations and lower first premolar agenesis less closely associated with PAX9 mutations.

Patients with oligodontia usually present associated craniofacial anomalies [11], in this case the patient did not show any underlying systemic condition [2]. Tome et al and Hartlev in 2019 and 2018 [3][5] respectively, report that the frequency of affected teeth is: lower second premolar, upper lateral incisor and upper second premolar and there is a significant correlation between the primary tooth and the absent successor. Donald et al in 2017 [4], report that the absence of canines is very rare, meanwhile in our report there is evidence of the absence of permanent second molars and lower central incisors, which leads to the need to report this case and make it known.

The sequelae of oligodontia include missing teeth, prolonged retention of primary teeth, root resorption and ankylosis resulting in unstable occlusion. The space for rehabilitation depends on the pattern and severity of tooth absence [2,3].

Early diagnosis by clinical and radiographic examination is essential [11,12]. Panoramic radiography is the most indicated for the diagnosis and study of dental absences in number, position of the teeth present and the state of the alveolar ridge. In addition, three-dimensional modalities such as 3D dental volumetric tomography and cone beam computed tomography can also provide complementary in-depth knowledge about the absence of dental germs [13].

On the other hand, Fenández et al in 2018 [14], mention that there is an association between the absence of teeth with the hypodivergent growth pattern coinciding with the case presented where the patient has a horizontal growth tendency, which could be explained by possible alterations in proliferation and development during odontogenesis [15]. Although Suyama et al in 2021 [9], found that patients with oligodontia showed Class III skeletal tendency with prognathism and flattened mandibular plane with a smaller gonial angle. These maxillofacial morphological characteristics may be induced by a deficiency in verticality and a statistically significant positive correlation between the higher number of missing teeth, Class III malocclusion, a decreased overjet. This would be due to which teeth are missing, upper or lower, because whenever treatment options are considered, skeletal growth areas should be respected. That is why the treatment of oligodontia could be a challenge because of the absence of teeth accompanied by a malocclusion where these conditions should be treated concomitantly and, in this case, oligodontia is presented in addition to a class II division 1 malocclusion.

Currently for this case a class 2 twin block appliance was placed to correct the malocclusion and a multidisciplinary therapy plan was carefully designed including orthodontics, dental implants, and esthetic restoration in the future.

Kang et al in 2016 [2], indicate that within the post-growth treatment options, spatial redistribution, prosthetic restoration with implant, through multidisciplinary treatments (pediatric dentistry, orthodontics, and oral rehabilitation) could be done. Optimal esthetics can be achieved by resolving the tooth size discrepancy between arches, gradually adjusting the gingival contour, and improving the crown width to length ratio, emphasizing that lifelong maintenance is essential for preservation [11].

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### 4. Conclusion

The absence of teeth together with a malocclusion is a challenge for the pediatric dentist and the multidisciplinary team to manage. Early diagnosis and treatment planning should be carried out to minimize the different complications

generated by these types of dental anomalies, performing a corrective management of the malocclusion concomitant with the management of spaces according to the pattern and severity of the oligodontia.

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## Compliance with ethical standards

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### *Disclosure of conflict of interest*

The authors declare that they have no known competing financial interests or personal relationships that may have influenced the work reported in this paper.

### *Statement of informed consent*

Informed consent was obtained from the participant included in the study.

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