VOLUME - 13, ISSUE - 07, JULY - 2024 • PRINT ISSN No. 2277 - 8160 • DOI : 10.36106/gjra

Original Research Paper

Oral Pathology

# NON- SYNDROMIC OLIGODONTIA WITH TAURODONTISM: AN ENIGMATIC CASE

| | |
|---|---|
| **Dr. Azra Kouser** | Post Graduate Student, Dept Of Oral Pathology & Microbiology, Indira Gandhi Govt Dental College, Jammu. |
| **Dr. Rubeena Anjum** | Professor And Head, Dept Of Oral Pathology & Microbiology, Indira Gandhi Govt Dental College, Jammu |
| **Dr Mandeep Kaur** | Assistant Prof., Dept Of Oral Pathology & Microbiology, Indira Gandhi Govt Dental College, Jammu. |

**ABSTRACT** Tooth agenesis is a frequently observed congenital anomaly in humans. While the absence of one or more teeth is common, the absence of multiple teeth is less frequent. Oligodontia refers to the congenital absence of six or more permanent teeth, excluding the third molars. Related terms like hypodontia or dental agenesis are used to describe the absence of primary or permanent teeth. The prevalence of oligodontia in permanent dentition is reported to be 0.14%. It typically occurs as part of a syndrome and rarely appears as an isolated condition. Genes responsible for non-syndromic oligodontia are found to be MSX1, PAX, AXIN2 and EDA gene, as well as environmental factors, are implicated in its etiology. Early diagnosis through clinical and radiographic examinations is essential for evaluating the number and position of present and absent teeth, as well as the condition of the alveolar ridge. This assessment is essential for comprehensive treatment planning involving a multidisciplinary team. Here we present an interesting case of oligodontia with taurodontism in a 14-year-old male child not associated with any syndrome.

**KEYWORDS :** Oligodontia; Hypodontia; Agenesis; Non-syndromic;

## INTRODUCTION

Tooth agenesis is the most common craniofacial congenital anomaly in humans. It is frequently identified during dental exams when one or more teeth are congenitally missing. While the complete absence of teeth is quite rare, the most prevalent forms are hypodontia and oligodontia. The condition is genetic in origin. Hypodontia involves the absence of 1 to 6 teeth whereas Oligodontia describes condition in which more than six teeth are missing [1].

The most commonly missing permanent teeth are the third molars (9-37%), followed by the mandibular second premolars (less than 3%), maxillary lateral incisors (less than 2%), and maxillary second premolars and mandibular incisors (less than 1% ) [2].

The precise cause of tooth agenesis is not fully understood, but genetic factors are widely recognized as significant contributors. Anomalies in the development of the mandibular symphysis, along with evolutionary trends leading to shortened dental arches, and disturbances in endocrine function are believed to impact the dental tissues responsible for forming tooth buds. These factors can result in the congenital absence of lower incisors [3].

Environmental factors have also been associated with tooth agenesis. These include localized infections of the jaw that could potentially damage tooth buds, disturbances in the endocrine system, systemic infections such as rubella, trauma to the apical region of the dentoalveolar process (caused by fractures or during extraction of deciduous teeth), exposure to certain chemicals or drugs like thalidomide or chemotherapy, radiation therapy, and disruptions in jaw innervation [4].

Together, genetic and environmental factors contribute to the complex etiology of tooth agenesis, influencing the development and absence of teeth [4].

Oligodontia may occur as a feature of specific diseases such as anhidrotic ectodermal dysplasia or incontinentia pigmenti, an X-linked genetic disorder that affects ectodermal structures and is associated with congenital tooth absence and abnormal form), Down's syndrome, Pierre Robin syndrome, and Ehler Danlos syndrome [5].

It can also present as an isolated condition (non -syndromic) and has been linked to mutations of MSX1, PAX9 and EDA genes [4].

There is a wide range of treatment options available, including removable and fixed prostheses as well as dental implants at a later age. Dental implants can be placed, restored and loaded in children suffering from syndromes like oligodontia and ectodermal dysplasia [6].

This paper presents a case of non-syndromic oligodontia, with agenesis of more than six teeth in the oral cavity along with taurodontism.

### Case Report

A 14 -year-old boy visited the Department of Oral medicine and radiology, concerned about his multiple missing teeth and spacing in his oral cavity since few years which his parent noticed and consulted the dentist, they were assured that it is normal till this age and advised to follow up every three months. There was no history of trauma or extraction, and both his medical and family histories were unremarkable.

Patient was well built and nourished with normal gait. Physical examination revealed no abnormalities in the nails or hair, normal perspiration, and no congenital cleft lip or palate. Extra oral examination revealed a face with normal facial profile and normal skeletal dental base relations [Figure 1]

**Figure 1:** Extra oral examination

Intraoral examination revealed mixed dentition stage, with

mesial step molar relationship, multiple unerupted permanent teeth, retained deciduous teeth, generalized interdental spacing and generalized stains.

The teeth present were all the four deciduous central incisors, maxillary right lateral incisors, all four deciduous canine, mandibular right and left permanent canine, two deciduous maxillary 1st molar, all four deciduous 2nd molar and permanent maxillary right and mandibular right and left 1st molar (Figure 2).

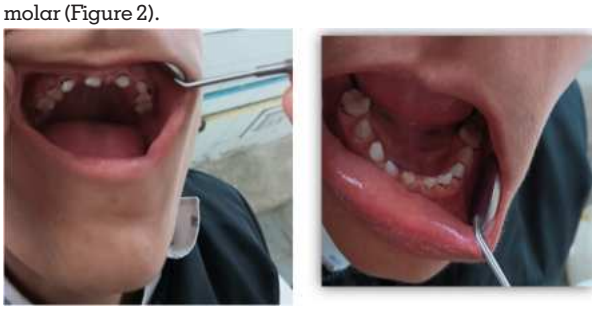

**Figure 2:** Intra oral examination showing retained deciduous teeth and interdental spacing

To ascertain the provisional diagnosis, an orthopantomography was advised which revealed no evidence of development of 11, 12, 13, 14,15, 17, 21, 22, 23, 24, 25, 26, 27, 31, 32, 34, 35,37 41, 42, 44, 45 and 47. Taurodontism was seen in relation to deciduous mandibular right 1st molar (Figure 3). Parents were informed about the agenesis of permanent teeth.

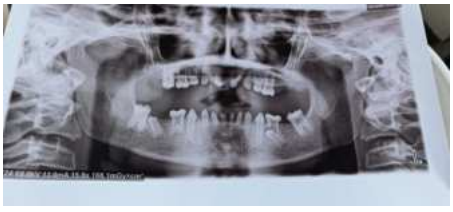

**Figure 3** OPG of the patient revealing retained deciduous teeth and agenesis of permanent teeth and taurodontism

Considering the history, clinical examination, radiographic finding a diagnosis of non-syndromic oligodontia with taurodontism was made. The treatment plan considered was reshaping of required teeth, extraction of retained deciduous teeth followed by removal/fixed partial denture for esthetic and functional rehabilitation of the patient.

To rule out any syndromic involvement, the patient was referred to the Department of Pediatrics. The medical evaluation revealed no presence of any syndrome.

## DISCUSSION

A tooth is considered congenitally missing if it cannot be detected clinically or radiographically, and there is no history of its extraction. In our case report, the fourteen -year-old child showed no signs of calcification of all permanent teeth except mandibular canines and maxillary right and mandibular right and left 1st molar, confirming the agenesis of these teeth [7]. Oligodontia is the term most commonly used to describe the phenomenon of congenitally missing teeth. It has been classified into two categories: isolated or non-syndromic, and syndromic hypodontia. Although oligodontia can occur over with 60 different syndromes, these anomalies can occur without any syndrome or systemic disease. However, oligodontia is seen more common in non-syndromic or familial form than syndromic form [8].

Syndromic oligodontia can occur as a dental trait due to mutations in genes such as PAX9, MSX1, AXIN2, EDA, and EDAR, or it can occur alongside systemic diseases like ectodermal dysplasia and Down syndrome, Nance-Horan syndrome, Rieger syndrome, Seckel syndrome, Wolf-Hirschhorn syndrome, Klippel-Feil syndrome, Van der Woude syndrome and cleft lip and palate [9].

The biological basis for the congenital absence of permanent teeth is partially explained by the failure of lingual or distal proliferation of tooth bud cells from the dental lamina. Hypodontia can be caused by environmental factors such as irradiation, tumors, trauma, hormonal influences, rubella, and thalidomide, as well as hereditary genetic dominant factors, or a combination of both. The MSX1 and PAX9 genes are crucial in early tooth development. PAX9, a paired domain transcription factor, plays a vital role in odontogenesis. To date, all identified mutations of PAX9 have been associated with the no syndromic form of tooth agenesis [10].

In this case report, oligodontia could not be associated with any syndrome as the thorough assessment by the pediatrician and Karyotyping did not reveal any significant findings. None of the family members suffered from a similar condition, suggesting the absence of a hereditary basis for the present defect. The prenatal and post-natal histories were noncontributory to suggest any environmental cause [3]. In addition to tooth agenesis, taurodontism was also seen in mandibular 1st molar.

The treatment options for using dental prostheses range from removable or fixed prostheses to dental implants, which can be considered once the period of active growth is complete. A long-term rehabilitation plan involving dental implants was discussed with the parents, but due to financial constraints, they opted for a more economical treatment option. Dental implants may be considered as a permanent solution at a later stage. In this case, the choice was between a removable or fixed prosthesis. Since the child was attending school and preferred not to have a removable prosthesis, the decision was made to fabricate a fixed prosthesis.

## CONCLUSION

The successful treatment of a patient with oligodontia and complex dentofacial abnormalities requires close and coordinated collaboration among an orthodontist, oral maxillofacial surgeon, pediatric dentist, and prosthodontist. Although oligodontia is often associated with several syndromes, the non-syndromic aspects should also be considered. It is important to treat the condition as early as possible to achieve both prosthetic and aesthetic functionality of the teeth.

**Financial support and sponsorship** -Nil.
**Conflicts of interest** -There are no conflicts of interest.

## REFERENCES

1. Tavajohi-Kermani H, Kapur R, Sciote JJ. Tooth agenesis and craniofacial morphology in an orthodontic population. *Am J Orthod Dentofacial Orthop* 2002; 122: 39-47
2. Grahen HJ. Hypodontia in permanent dentition. Odontol Rev 1956;7:1.
3. De Coster PJ, Marks LA, Martens LC, Huysseune. Dental agenesis: genetic and clinical perspectives. J Oral Pathol Med 2009; 38:1-17.
4. P. Pannu, V. Galhotra, P. Ahluwalai, R. S. Gambhir. Non-Syndromic Oligodontia In Permanent Dentition: A Case Report. Ghana Medical Journal September 2014 Volume 48, Number 3.
5. Kurisu K, Tabata MJ. Review. Human genes for dental anomalies. Oral Diseases 1997;3:223-8.
6. Kramer FJ, Baethge C, Tschernitschek H. Implants in children with ectodermal dysplasia: a case report and literature review. Clin Oral Implants Res 2007;18(1):140-6
7. Kjaer I, Kcosis G, Nodal M, Christensen LR. Aetiological aspects of mandibular tooth agenesisfocusing on the role of nerve, oral mucosa, and supporting tissues. Eur J Orthod 1994; 16(5):371- 5.
8. Cobourne MT. Familial human hypodontiais it all in the genes? *Br Dent J*, 2007; 203:203-208.
9. Bustos Castellano Andrea Tatiana 1 * and Ochoa Bermeo María Fernanda Non-syndromic oligodontia: A case report. World Journal of Advanced Research and Reviews, 2022, 15(02), 472–477
10. Guruprasad R, Nair PP, Hegde K, Singh M. Case report: nonsyndromic oligodontia. JIDA, 2011; 3:450-454