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CASE REPORT

Oligodontia: A Case Report and Review of Literature

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ABSTRACT

Oligodontia, a form of hypodontia commonly seen in permanent than in deciduous dentition. It is defined as the congenital absence of six or more teeth, excluding third molars. It is relatively a rare condition that can occur either as an isolated finding or as part of a syndrome. A case of nonsyndromic oligodontia in a 12-year-old boy with congenital absence of all permanent teeth except the premolars (excluding third molars) and over-retained deciduous dentition with conical shaped anteriors is reported. The prevalence, possible etiological factors and treatment options of the condition is reviewed.

Keywords: Oligodontia, Hypodontia, Congenital absence of teeth, Dental agenesis.

INTRODUCTION

Dental agenesis is the most common developmental anomaly in humans, often presenting a significant clinical problem.[1] It is classified according to the number of missing permanent teeth, excluding the third molars. Hypodontia refers to a condition with one to five missing teeth. Patients with six or more missing teeth are classified as having oligodontia; anodontia is the term given to the complete absence of teeth.[2] The incidence of missing permanent teeth (excluding third molars) has been reported to vary from 2.6 to 11.3% depending on demographic and geographic profiles, whereas the incidence of missing primary teeth is considerably low.[1] Oligodontia is relatively a rare condition, probably affecting about 0.1 to 0.2% of the population.[3] It may occur as a part of a syndrome[4] or as an isolated condition that has been linked to mutations of the MSX1 and PAX9.[2]

Oligodontia has a wide variety of manifestations.[5] Depending on the number and location of missing teeth, masticatory, speech and esthetic problems may arise.[5] Different positional changes of teeth, their morphology and size may occur simultaneously with oligodontia. It is also associated with growth disturbances of the maxillofacial skeleton, and thus the facial appearance.[3] The treatment of such patients, therefore, is often complex.[3] In addition to loss of function and esthetic compromise, psychological development is an important concern in the oral rehabilitation of growing patients. Although oligodontia is a rare congenital disorder, treatment for this abnormality can be a challenge.[6] The main goal of management is to improve esthetics, mastication and speech.[4] Care often requires multidisciplinary approach. The purpose of this article is to report a rare case of congenitally missing all permanent teeth except the premolar buds (excluding third molars) and over-retained deciduous dentition with conical shaped anterior teeth and review the literature.

CASE REPORT

A 12-year-old boy visited the Department of Oral and Maxillofacial Pathology along with his parents with the chief complaint of abnormally shaped teeth. The detailed history revealed that he was teased of having crocodile teeth by his school friends. Extraoral examination revealed facial symmetry and no skeletal changes. Intraoral examination revealed presence of all deciduous teeth without any permanent tooth. Over-retained deciduous teeth were present. Retained deciduous anteriors were conical in shape and showed incisal attrition. He also had an edge-to-edge bite (Fig. 1). Panoramic radiograph revealed the absence of all permanent teeth except the buds of developing premolars, along with conical shaped retained primary teeth. There was no evidence of root resorption with retained primary teeth (Fig. 2). Deciduous mandibular molars bilaterally exhibited mesotaurodontism (Figs 3A and B). Family history was noncontributory. Thorough physical evaluation of the patient did not reveal any other systemic abnormalities. Based on clinical and radiological examinations, diagnosis of nonsyndromic oligodontia was made. The condition was explained to the patient and parents and treatment plan was discussed. However, patient's parents were reluctant for their child's treatment.

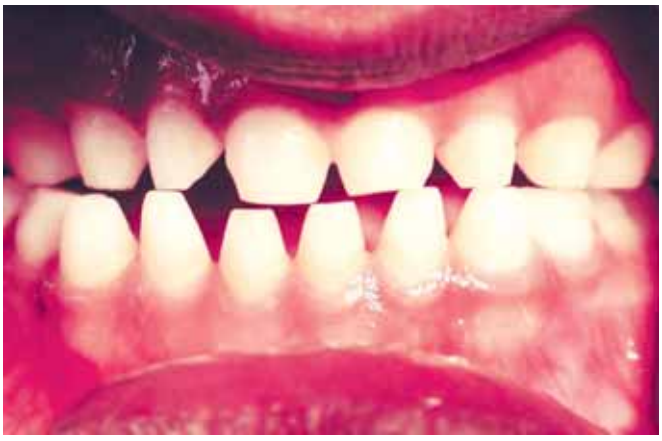


Fig. 1: Clinical photograph showing conical shaped deciduous anteriors with incisal attrition

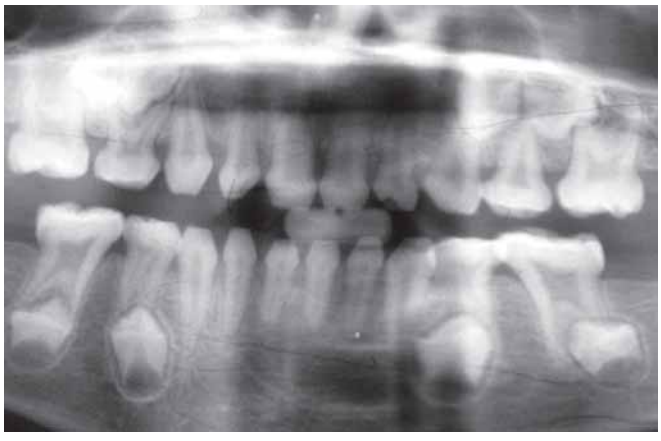


Fig. 2: Orthopantomograph (OPG) exhibiting retained deciduous teeth without root resorption and presence of premolar buds only

REVIEW OF LITERATURE

Hypodontia or tooth agenesis is defined as a common anomaly of human dentition characterized by the developmental absence of one or more teeth.[7,8] Oligodontia is defined as congenital absence of 'many' teeth, without any specification as to the meaning of 'many.[9] It is also seen that the term oligodontia is used to define developmental absence of multiple teeth, usually associated with systemic manifestations.[10]

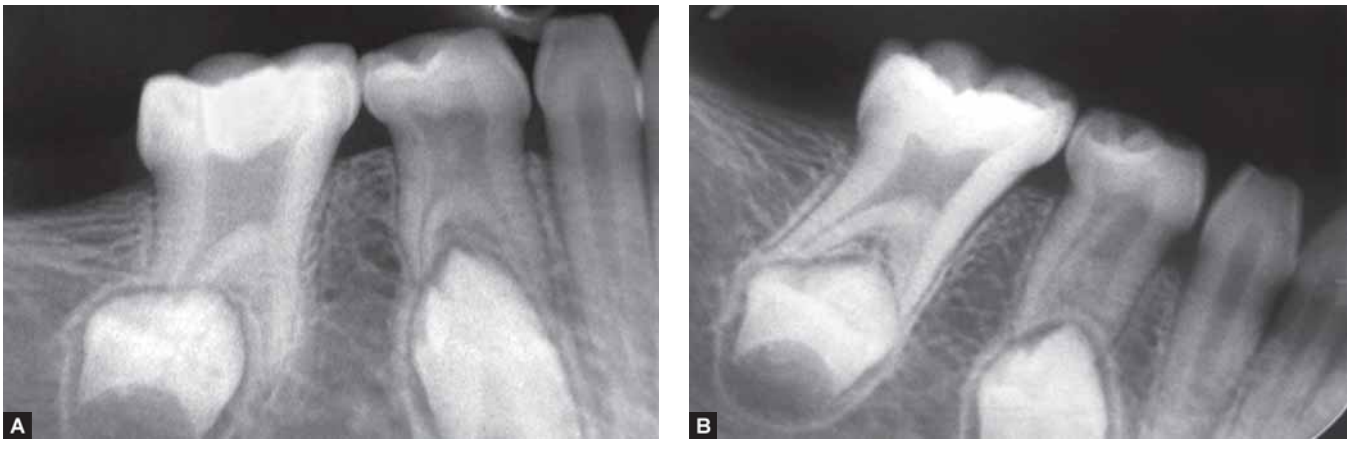
Schalk-van der Weide has suggested six or more permanent teeth (excluding third molars) as a criterion.[3,9] According to a 1996 Consensus Conference on Oral Implants in Young Patients, the following definitions are used: 'Hypodontia' is defined as the absence of one to five permanent teeth, while the term 'oligodontia' refers to the absence of six or more permanent teeth and 'anodontia' to the absence of all permanent teeth.[11]

Thus, oligodontia is generally defined as the congenital absence of six or more permanent teeth, excluding the third molars.[5,8,12] It is also known as partial anodontia, severe or advanced hypodontia.[8,13] Some of them also refer to this as selective tooth agenesis.[14]

Population studies on the prevalence of hypodontia suggest that majority (more than 80%) will present with one or two congenitally missing teeth and only less than 1% will present with six or more.[15] Oligodontia represents a relatively rare condition.[3] The incidence of oligodontia is reported to vary from 0.08 to 0.16%.[2] Rolling S reported only 0.16% of the Danish school children with five or more missing permanent teeth.[3,9] In a recently published study, a rate of 0.16% oligodontia was reported.[3] The incidence rate suggested by Schalk-van der is 0.08% of the population.[3,11] In the study by Ravan and Nielsen of 1530 Danish school children, 0.26% had six or more missing teeth. In a Swedish study prevalence was 0.19%.[3] Its prevalence is estimated at 0.14% in the white population.[16]

Frequency of congenitally missing teeth was higher in girls than in boys in one of the studies on oligodontia patients. However, in another study there was no significant difference.[8] Data from the Danish school children study showed that condition is more frequent in girls than in boys.[9]

The pattern of tooth absence is influenced by the gene affected, as well as the type of mutation within the specific gene.[2] It affects permanent rather than deciduous dentition.[14] The teeth most commonly absent are the permanent second premolars and the maxillary lateral incisors.[2] The permanent fist molars and maxillary central incisor are the most stable teeth.[2,3] Molar absence, however, is a prominent feature of some forms of isolated oligodontia.[2]



Figs 3A and B: Intraoral periapical radiograph (Iopa) showing mesotaurodontism with deciduous mandibular molars

Oligodontia can occur in association with genetic syndromes, such as ectodermal dysplasia, incontinentia pigmenti, Down syndrome and Rieger syndrome[2,5] or as a nonsyndromic isolated familial trait, or as a sporadic finding.[17] It is also reported that it may be a common (even though previously not well documented) feature in Wolf-Hirschhorn syndrome.[18] When oligodontia is associated with a syndrome there may be abnormalities of the skin, nails, eyes, ears and skeleton.[4]

Severe hypodontia or oligodontia is often associated with conical teeth, microdontia, and delayed eruption of permanent teeth, an increased free way space and retention of deciduous teeth.[4,19] Tooth agenesis has also been reported in association with taurodontism. The studies that suggested association between tooth agenesis and taurodontism imply that the association is more likely with severe tooth agenesis or oligodontia.[20]

Dentoalveolar characteristics associated with oligodontia include occlusal disturbances like deep bite, cross bite, attrition, steep inclination of maxillary incisors, disturbances of eruption—delayed eruption, ectopic eruption and alterations of tooth morphology, i.e. microdontia, conical shape of incisors and canines.[3]

Oligodontia is also associated with disturbances of the maxillofacial skeleton.[3] Characteristic growth disturbances and changes of facial appearance include maxillary retrognathism and hypoplasia, mandibular retrognathism, decreased vertical and transverse dimensions of alveolar process, lower anterior facial height, receding midrace, and deep labial-mental fold.[3]

Oligodontia is a heterogenous condition. Patients with oligodontia can be classified into three different types according to their clinical presentation and complexity of their prosthodontic requirements.[2]

Recent advances in the human genetics and molecular biology are providing us with a greater understanding of tooth development.[15] Much information on the genes and transcription factors that regulate odontogenesis has been obtained from murine tooth model and transgenic mice.[1] Previous studies have shown that some genes have a strong influence on tooth development (MSX1, PAX9, LEF1, PITX2) whereas other genes have a less pronounced effect (DLX1, DLX2, GLI2, GLI3).[1]

Abnormal gene function may also disrupt specific signalling pathways involved in tooth development, resulting not only in abnormal tooth number but also in abnormal tooth size and or shape.[1] Several genes and disruptions in the molecular pathways are suggested in causing defects affecting all teeth in a majority of cases presenting as oligodontia or anodontia.[1] List of the genes and molecular pathways involved in tooth agenesis are shown in Table 1.

Dental agenesis may also result from failure of [1] FGFs—FGF-4, -8,-9, -20 in the epithelium, FGF3, -7, and -10 in the mesenchyme, BMPs—BMP-2, -4, -7 in the epithelium and BMP-2 through -7 in the mesenchyme and their respective receptors required as targets or in feedback mechanisms during early tooth development.[1]

Table1: List of the genes and molecular pathways involved in tooth agenesis[1]

- Wnt/b–catenin/LEF 1
- MSX1
- MSX2
- SHH
- p63
- Pitx2
- Runx2/Cbfa1

In the last decade, more light has been shed on the multifactorial etiology of oligodontia.[5] Endocrine, local, environmental and hereditary factors of congenitally missing teeth have been suggested and identified, the latter through molecular genetics.[5]

Nonsyndromic familial oligodontia has, in most cases, been shown to be inherited as an autosomal dominant trait.[17] Mutations in the genes MSX1 and PAX9 that encode transcription factors were demonstrated to be associated with isolated, nonsyndromic oligodontia.[5] LTBP3 is the third gene which is identified as causing oligodontia or selective tooth agenesis.[14] Furthermore, agenesis of few teeth to entire set of teeth in one large Chinese kindred has been mapped to locus on chromosome 10q11.[17] The literature reports eight families in whom PAX9 mutations segregate with nonsyndromic autosomal-dominant inherited oligodontia.[17] A frameshift mutation recently identified within the paired domain of the transcription factor, PAX9, has been linked to a unique form of oligodontia in a single multigenerational family.[7] Severe case of nonsyndromic oligontia was associated with a large heterozygous deletion on chromosome14, which included the whole PAX9 gene.[17]

The genes for two syndromes involving hypodontia have been identified. The EDA gene encodes a transmembrane protein that is involved in hypohydrotic/anhydrotic ectodermal dysplasia. The RIEG gene causes Rieger syndrome an autosomal dominant condition that involves missing teeth, anomalies of eyes and umbilicus.[7]

Because of its variable genetic etiology, the presentation and subsequent clinical effect on the dentofacial structures of patients diagnosed with oligodontia may vary greatly.[2] Congenital missing teeth can create dental and facial disfigurement, which can lead to social withdrawal, especially in adolescent years.[6] Prosthodontic treatment of oligodontia patients is, therefore, important for functional, esthetic and psychological reasons.[21] Treatment of such patients requires a fully integrated interdisciplinary team approach of orthodontists, oral and maxillofacial surgeons and prothodontists.[3] Treatment options depend on the severity of the condition and patient's perceived need for care.[21] A number of factors must be taken into account at the time of treatment planning, which include age of the patient, number and condition of retained teeth,

number of missing teeth, condition of supporting tissues, the occlusion and the interocclusal space.[10] Common methods of treatment employed include, prerestorative orthodontics which is frequently required to move teeth to a favorable position, restoration with removable and fixed partial dentures and restoration with implant-supported prosthesis.[10,21] Increased experience with oral implants and supplementary augmentation techniques have created new options for treatment of patients with oligodontia.[11]

In the present report, the boy presented with congenital absence of all permanent teeth except the premolars (excluding third molars) and over-retained deciduous dentition. Over-retained deciduous anterior teeth were conical in shape. Psychosocial distress was evident in the present case because of the presence of abnormal shaped teeth. Thorough evaluation did not reveal any significant family history and systemic abnormalities. However, genetic evaluation was not carried out. Though the patient and his parents were concerned about the esthetics, they were hesitant to take the treatment. Due to lack of cooperation, treatment was not performed. Such an approach becomes a major hindrance to successful treatment of such cases.

CONCLUSION

Current understanding of rare conditions like oligodontia may be enhanced by reporting of such cases. Patients with oligodontia may not only have functional problems but also psychological distress which requires early diagnosis and proper evaluation to improve oral health and psyche of the child.

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