

# Nonsyndromic oligodontia: A rare case report

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## ABSTRACT

Oligodontia is generally defined as agenesis of six or more teeth excluding the third molars. It is a rare finding which has not been frequently documented in Indian children. Most often oligodontia appears as a part of some congenital syndromes. The exact etiology for oligodontia is unknown. The consequences of missing teeth include abnormal occlusion or altered facial appearance, which can cause psychological distress in some patients.

**Key words:** Agenesis, ectodermal dysplasia, hypodontia

## INTRODUCTION

One of the most common anomalies seen in the permanent dentition is the agenesis of one or more teeth. Various terminologies have been used to describe the agenesis of teeth in the primary or permanent dentition. Hypodontia is used to describe the agenesis of one or few teeth; Oligodontia is used to describe agenesis of six or more teeth excluding the third molars. Anodontia is the extreme of oligodontia where there is total absence of any dental structure.<sup>[1]</sup> Hypodontia has a prevalence of 1.6-9.6% in the permanent dentition, excluding agenesis of the third molars. Oligodontia has a population prevalence of 0.3% in the permanent dentition.<sup>[2]</sup> Oligodontia occurs more frequently in girls in a ratio of 3:2. Agenesis of only the third molars has a prevalence of 9-37%. In the deciduous dentition, hypodontia occurs less often (0.1-0.9%) and has no significant sex distribution.<sup>[2,3]</sup> The mandibular second premolar is the most frequently missing

tooth after the third molar, followed by the maxillary lateral incisor and the maxillary second premolar. Agenesis of maxillary central incisors, canines, or first permanent molars seems to be rather exceptional.<sup>[4]</sup>

Hypodontia and oligodontia are classified as isolated or nonsyndromic hypodontia/oligodontia and syndromic hypodontia/oligodontia or hypodontia/oligodontia associated with syndromes. Oligodontia is often associated with specific syndromes and/or severe systemic abnormalities, while anodontia is commonly seen in severe cases of ectodermal dysplasia.<sup>[3]</sup> Congenital absence of teeth may be due to physical obstruction or disruption of the dental lamina, space limitation, and functional abnormalities of the dental epithelium or failure of initiation of the underlying mesenchyme. Characteristic dental symptoms are a reduced number of teeth, a reduction in tooth size, anomalies of tooth form, and delayed eruption. The absence of teeth in young patients can cause esthetic, functional, and psychological problems particularly if the teeth of the anterior region are involved. The purpose of this article is to report a rare case of nonsyndromic oligodontia with agenesis of 11 permanent teeth excluding the third molars in a 9-year-old patient. The diagnosis of hypodontia/oligodontia should be performed as early as possible in order to prevent esthetic and functional problems in dentition.<sup>[3]</sup>

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### Definition/diagnosis criteria

A tooth is defined as congenitally missing if it has not erupted in the oral cavity and is not visible in the radiograph. All primary teeth have erupted by the age of 3 years and all permanent teeth except third molars between the ages of 12 and 14 years. Therefore 3-4-year-old children are suitable for

diagnosis of congenitally missing primary teeth by clinical examination and 12-14-year-old children, for diagnosis of permanent teeth excluding the third molars.<sup>[4]</sup> The use of panoramic radiography is recommended, together with clinical examination for the detection or confirmation of dental development and performing the diagnosis of hypodontia.<sup>[5]</sup>

## CASE REPORT

A 9-year-old female patient reported to the department of pediatric dentistry with a chief complaint of irregularly placed upper front teeth for past 3 years. The patient's past medical history and the family history were not significant. It was patient's first visit to a dentist. There was no abnormality detected on general examination. Extraoral examination revealed no abnormalities of the skin, hair, or nails [Figure 1]. On intraoral examination, patient was having mixed dentition with retained deciduous teeth 71, 81 [Figures 2 and 3]. The orthopantomographic (OPG) examination revealed agenesis of 15 permanent teeth including third molars. The missing teeth were # 13, 15, 16, 17, 18, 23, 25, 26, 27, 28, 31, 37, 38, 41, and 48 [Figure 4].

The patient was diagnosed as a case of oligodontia, since more than six permanent teeth were congenitally missing excluding the third molars.

In view of the oligodontia of permanent teeth, the patient was referred to a pediatrician to rule out any associated syndromes and systemic disorders. A detailed examination was done to rule out abnormalities associated with the skull, chest, vertebrae, and clavicles. The patient was normal and did not show any physical or skeletal abnormality. The ophthalmological, dermatological, and neurological examination of the patient revealed no pathological symptoms and showed no signs of mental retardation. Based on the above findings the case was finally diagnosed as nonsyndromic oligodontia of 11 permanent teeth excluding the third molars. However, the genetic evaluations were not performed.

## DISCUSSION

Hypodontia/oligodontia is an anomaly that may result in dental malpositioning; periodontal damage; lack of



Figure 1. Extraoral examination



Figure 2. Clinical photograph of maxillary arch showing missing teeth



Figure 3. Clinical photograph of mandibular arch



Figure 4. Orthopantomogram (OPG) showing congenital absence of multiple permanent teeth and retained primary teeth

development of maxillary and mandibular bone height; and has significant psychological, esthetic, and functional consequences.<sup>[3]</sup> Oligodontia can occur in association with various genetic syndromes, such as ectodermal dysplasia, incontinentia pigmenti, Down syndrome, Rieger syndrome, Wolf–Hirschhorn syndrome, Van der Woude syndrome, ectrodactyly-ectodermal dysplasia-clefting syndrome, cleft lip palate ectodermal dysplasia syndrome, orofacial digital syndrome type I, Witkop's tooth and nail syndrome, Fried syndrome, hair-nail-skin-teeth dysplasias, Hirschhorn syndrome, hemifacial microsomia, and recessive incisor hypodontia.<sup>[3]</sup> When oligodontia is associated with a syndrome, there may be abnormalities of the skin, nails, eyes, ears, and skeleton. The exact etiology for oligodontia is unknown. Various factors have been described in the literature. Heredity is the main etiological factor, several environmental factors like viral infections, toxins, and radiotherapy or chemotherapy may cause agenesis of permanent teeth. However, most of the cases are caused by genetic factors. The heritability of congenitally missing teeth has been shown in many studies. The genetic factors may be dominant or recessive and it is obvious that in many cases multiple genetic and environmental factors are acting together. The importance of genetic factors is shown by appearance of multiple cases among relatives and higher concordance in identical than in nonidentical twins. It is also reported that several genes, which when defective, cause congenitally missing teeth. Mutations of MSX-1 and PAX-9 genes have been associated with agenesis of teeth.<sup>[3]</sup> Mutations in different genes may cause different types of oligodontia, that is, different sets of teeth are missing. Perhaps the best family study of tooth agenesis was done by Grahnen in 1956. He found that if either parent had one or more congenitally missing teeth, there was an increased likelihood that their children would also be affected. This familial relationship suggests that the genes are important.<sup>[6]</sup>

Comprehensive literature review shows only countable number of cases reporting maximum number of congenitally missing of permanent teeth. Tsai *et al.*, have reported a case of oligodontia in a 6-year-old female patient with congenital absence of 16 permanent teeth.<sup>[7]</sup> Akhya *et al.*, in their case report of 16-year-old patient have reported oligodontia of six permanent teeth.<sup>[8]</sup> Rasmussen reported nine cases of nonsyndromic oligodontia with 14-24 missing teeth excluding third molar.<sup>[9]</sup> Nagveni *et al.*, have reported a case of nonsyndromic oligodontia in a 13-year-old patient with 14 missing teeth.<sup>[10]</sup> Guruprasad *et al.*, have reported a case of nonsyndromic oligodontia in a 9-year-old patient with 26 missing teeth. The present case report shows developmental agenesis of 11 permanent teeth excluding the third molars with no identifiable etiology. Oligodontia was not associated with family history and the patient was not suffering from any syndromes or systemic disorders.

Oligodontia and hypodontia have similar associated anomalies. Several dental anomalies have been reported together with congenitally missing teeth. Examples of these are delayed formation and eruption of teeth, reduction in tooth size and form, malposition of teeth (ectopic maxillary canines and ectopic eruption of other teeth), infraposition of primary molars, teeth with short roots, taurodontism, rotation of premolars and or maxillary lateral incisors, enamel hypoplasia, hypocalcification, and dentinogenesis imperfecta.<sup>[3]</sup> However, in the case reported here, the remaining teeth were normal in color, size, and shape.

Patients suffering from oligodontia may have severe psychological, esthetic, and functional problems.<sup>[3]</sup> Thus, early diagnosis and treatment of these patients is very important. The treatment of oligodontia could be challenging if there are several missing teeth and malocclusion is present.<sup>[10]</sup> A number of factors must be taken into account for success of the treatment, important being the age of the patient. There are a number of options available to restore space generated by missing teeth. Dental treatment can vary depending on the severity of the disease and generally requires a multidisciplinary approach. Treatment options include orthodontic therapy, implants, adhesive techniques, removable partial prostheses, fixed prostheses, and overdentures. Most cases require the construction of a partial denture as an intermediate procedure before fixed prostheses are constructed.<sup>[10]</sup> Treatment not only improves speech and masticatory function but also has psychological implications that may greatly help in regaining self-confidence of the patient.

## CONCLUSION

Current understanding of rare conditions like oligodontia may be enhanced by reporting of such cases. Oligodontia cases should be evaluated carefully for the presence of any syndromes and managed appropriately. Patients suffering from oligodontia may have severe functional, esthetic, and psychological problems. Hence, the management of such patients generally requires a multidisciplinary approach.

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