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Non-syndromic Oligodontia in Permanent Dentition of Monozygotic Twins: Report of a Rare Case

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Abstract

Aim: To report a case of non-syndromic oligodontia involving 26 permanent teeth in monozygotic twin sisters and to discuss the possible genetic etiology, inheritance pattern and associated dental anomalies of this condition.

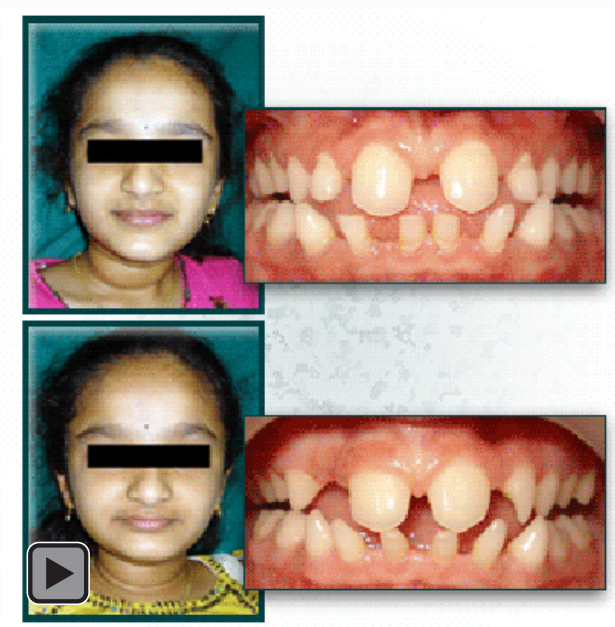
Background: Hypodontia constitutes one of the most common developmental anomalies in humans and is defined as developmental absence of one or more teeth with reported prevalence of 1.6 to 9.6% in the permanent dentition. Oligodontia is defined as agenesis of six or more teeth excluding third molars.

Case Report: This article describes a case of non-syndromic oligodontia involving 26 permanent teeth in monozygotic twin sisters. The twins had positive family history of hypodontia in their paternal grandmother, parental consanguinity (first cousin) and similarity in pattern of oligodontia. Root formation of permanent maxillary first molars and central incisors was delayed in both the twins.

Summary: This article reports a case of non-syndromic oligodontia in permanent dentition of monozygotic twins. Possible genetic etiology, inheritance pattern and associated dental anomalies are discussed.

Clinical Significance: Strong genetic link associated with oligodontia help the dentist to know the possibility of its occurrence in other family members and in future generations.

Keywords: Oligodontia, monozygotic twins, hypodontia, genetic consanguinity.



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Introduction

Hypodontia constitutes one of the most common developmental anomalies in humans and is defined as developmental absence of one or more teeth from the dentition.[1,2] Other frequently used terms are *oligodontia*, *partial anodontia*, and *anodontia*. Oligodontia is defined as agenesis of six or more teeth excluding third molars.[3] Anodontia, the total

absence of teeth, is rare, and usually associated with hypohidrotic ectodermal dysplasia.[1] The term *partial anodontia* is contradictory and no longer used.[1,4] The commonly used term *congenitally missing teeth* is a misnomer as teeth are absent in the mouth at birth. Tooth agenesis is a more informative term as it implies the underlying developmental defect.[5] Oligodontia is classified as isolated or nonsyndromic oligodontia and syndromic oligodontia.[6]

Hypodontia may occur in association with other genetic diseases, or as an isolated familial or sporadic form, and frequently affects the permanent rather than the primary dentition. The prevalence in the primary dentition is less than 1% of the population and frequently involves the mandibular incisors.[1,2]

The teeth most frequently affected in the permanent dentition are the third molars, followed by lateral incisors and second premolars.[1,7] Reported prevalence of hypodontia in the permanent dentition varies from 1.6% to 9.6% in the population, excluding third molars, which occur in up to 25% of the population.[7]

Early recognition of oligodontia is vital to provide adequate treatment and prevent malocclusion. Oligodontia may have significant psychological, esthetic, and functional consequences. Treatment of patients with oligodontia generally requires a multidisciplinary approach. Options for treatment depend upon the age of the patient and the severity of oligodontia and require an integrated approach involving pediatric, orthodontic, and restorative skills. Treatment can include space closure or space opening before restorative procedures with orthodontic therapy, removable or fixed partial dentures, implant-retained prosthesis, or a combination of these treatment strategies.

This article describes monozygotic twin sisters with nonsyndromic oligodontia involving 26 permanent teeth. To the best of our knowledge, no such cases have been reported in the literature.

Case Report

Twelve-year-old twin sisters reported with a complaint of failure of shedding of deciduous teeth and in eruption of permanent teeth. Their parents, who had normal dentition, had a consanguineous marriage (first-cousin marriage) and the paternal grandmother reportedly had a similar problem of over-retained deciduous teeth and missing permanent teeth. Both the girls had attained their developmental milestones for their age except for the dentition. Their prenatal and natal histories were uneventful and their mother gave no history of exposure to radiation or any medications during pregnancy. No history of orofacial trauma or unusual childhood diseases was reported. No systemic disease or syndrome was determined. The twins had identical features (Figure 1A and B). Their skin and hair appeared normal. Their height and weight were within normal limits.

On dental examination, Twin I revealed the presence of retained deciduous teeth: right maxillary lateral incisor to second molar, left

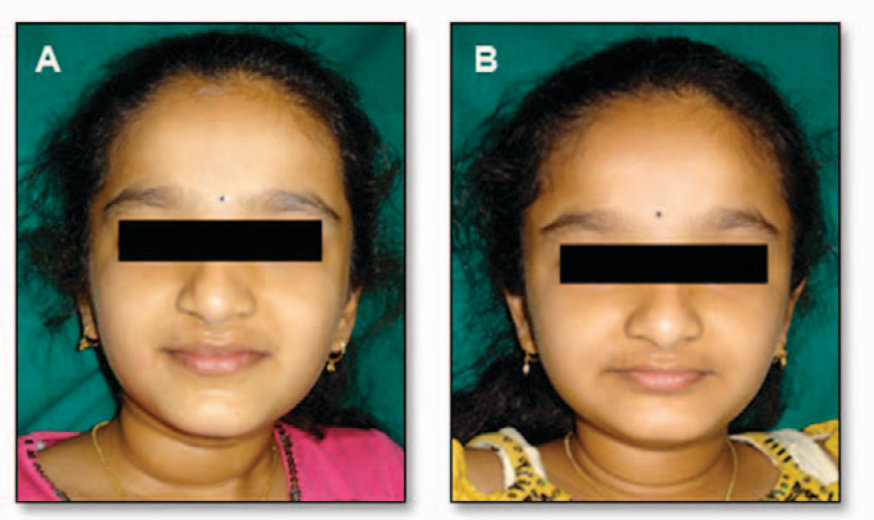

Figure 1. A and B. Facial photographs of twins I and II showing identical features.

maxillary lateral incisor to second molar, and right and left mandibular central incisor to second molar. Erupted/erupting permanent teeth were maxillary right and left central incisors, maxillary right and left first permanent molars, and (erupting) right mandibular second permanent molar (Figures 2A, 3A, 4A).

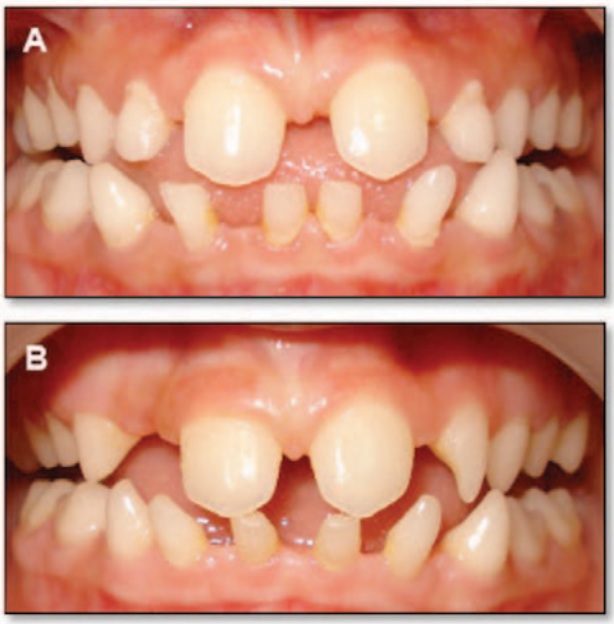

Figure 2. A and B. Intraoral photograph showing buccal view of the dentition of twins I and II respectively.

Twin II revealed the presence of retained deciduous teeth: right maxillary canine to second molar, left maxillary lateral incisor to second molar, and right and left mandibular central incisor to second molar. Erupted permanent teeth were maxillary right and left central incisors and maxillary right and left first permanent molars (Figures 2B, 3B, 4B).

Both the twins had cone shaped crowns of mandibular primary incisors and morphologically altered crowns of permanent maxillary central incisors. Oral mucosa of both the twins appeared normal.

Panoramic radiographs (Figure 5A and B) of both cases revealed bilateral absence of all the permanent teeth that were clinically missing except right and left mandibular second permanent molars. Root formation of maxillary first molars and central incisors was not completed. Maxillary first permanent molars were single-rooted. There was no significant resorption of roots of the retained deciduous teeth except the first molars. Bone of the maxilla and the mandible showed normal aspect.

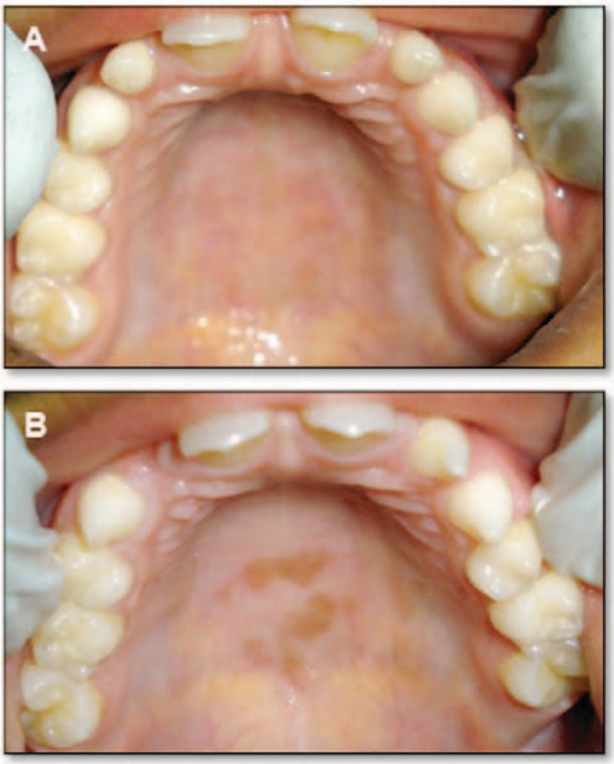

Figure 3. A and B. Intraoral photograph showing upper arch of twins I and II respectively.

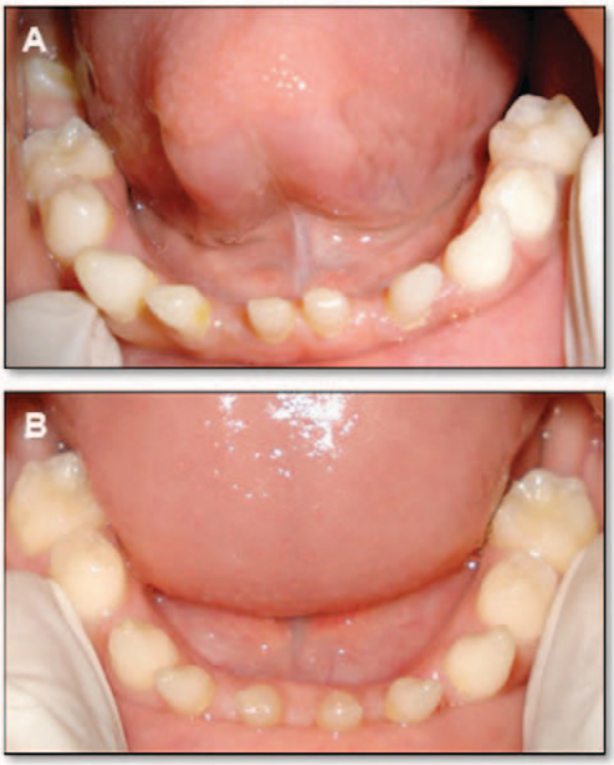

Figure 4. A and B. Intraoral photograph showing lower arch of twins I and II respectively.

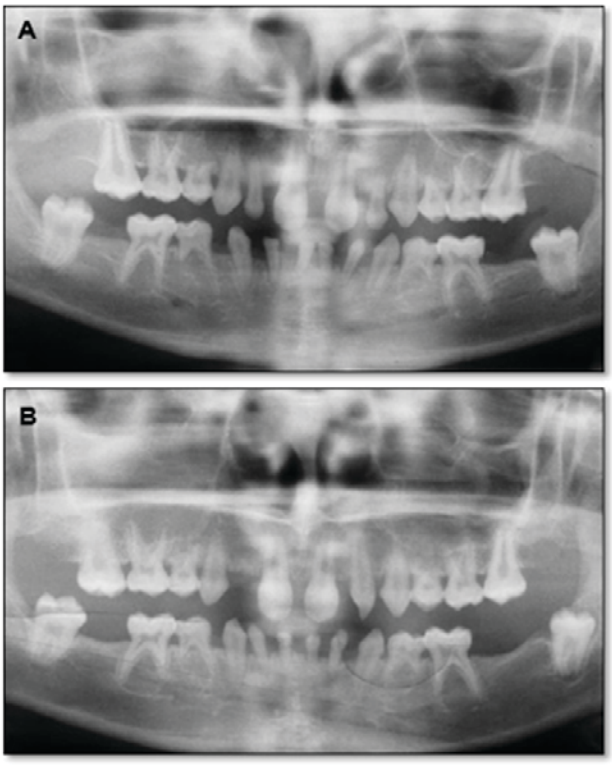

Figure 5. A and B. Cropped image of panoramic view of twins I and II respectively.

Considering the history and the clinical and radiographic findings, a diagnosis of monozygotic twin sisters with nonsyndromic oligodontia was arrived at.

The condition was explained to parents and pedodontic consultation was sought. No active treatment was instituted at the first visit as the retained deciduous teeth did not show significant root resorption. Patients are under regular follow up at our hospital and treatment will be instituted as and when required.

Discussion

Studies of twins and multiple births have proved that oligodontia is a heritable trait.[7] Our cases had positive family history of hypodontia in their paternal grandmother, parental consanguinity (first cousins), and similarity in pattern of oligodontia. This confirms the genetic etiology of hypodontia. However, an associated syndrome was absent.

Many investigators have suggested a multifactorial etiology of oligodontia, combining polygenic and environmental influences.[5,8–10] Although tooth agenesis is occasionally caused by environmental factors, in the majority of cases hypodontia has a genetic basis. Developing teeth are affected by environmental factors such as multiagent chemotherapy, radiation therapy, fractures, surgical procedures on the jaws, extraction of the preceding primary teeth, and lack of necessary space imposed by malformed jaws.[6] However, none of these environmental factors attributed to oligodontia in our case. An evolutionary trend towards fewer teeth has been proposed as a contributing factor in hypodontia.[7]

In familial hypodontia, the type of inheritance in the majority of families seems to be autosomal dominant with incomplete penetration and variable expressivity.[5] An autosomal recessive model of inheritance is also possible.[5] Mutations in transcription factors *MSX1* and *PAX9* have been identified in families with an autosomal dominant oligodontia.[3]

The most distinguishing feature of *MSX1*-associated oligodontia is the frequent (75%) absence of maxillary first bicuspids, while the most distinguishing feature of *PAX9*-associated oligodontia is the frequent (>80%) absence of the maxillary and mandibular second molars.[11] Our cases showed the absence of maxillary second permanent molars and maxillary bicuspids.

Recently, a Finnish family with dominantly transmitted oligodontia and colorectal polyps was shown to carry a germline *R656X* mutation in *AXIN2*.[12] Hence cases of oligodontia should be screened and followed for colorectal neoplasia.

Hattab[13] had reported oligodontia in two sisters with polycystic ovarian syndrome (PCOS). They collectively had 56 developmentally missing permanent teeth. However, our cases were not evaluated for PCOS as they were young and had not attained menarche.

Patients with hypodontia show a tendency for delayed tooth formation.[14] In our cases, root formation of maxillary first molars and maxillary central incisors was delayed.

It is interesting to note that maxillary central incisors (the most stable teeth in addition to the first molars), which are frequently missing teeth in patients with ectodermal dysplasia, are present in our cases.

A strong genetic link associated with oligodontia should alert the clinician about the importance of family history in these cases and also help the clinician to know the possibility of its occurrence in other family members and in future generations. Also, the detailed history including prenatal and natal history, orofacial trauma, and medical history is important to rule out environmental etiologic factors of hypodontia and any systemic syndromes.

Panoramic radiography is a useful diagnostic tool for the diagnosis of oligodontia. These provide a global view of the jaws not only for diagnosing oligodontia but also for evaluating other anomalies of the teeth such as morphologic alterations and variations of tooth size.[15] Avcu[16] recommended a panoramic radiographic examination when a tooth was missing because it might be an ectopic impaction. In the present cases panoramic radiographs revealed bilateral absence of all the permanent teeth that were clinically missing except for the mandibular second permanent molars. The radiograph also revealed single-rooted maxillary first molars while evaluating the image for other possible developmental abnormalities.

The principal aim of treatment in cases of hypodontia is to replace the missing teeth and hence improve the patient's appearance, speech, and masticatory efficiency. Absence of a moderate number of teeth may be managed with fixed or removable prostheses, but larger tissue deficiencies within the arches may usually be handled only with removable prostheses.[17]

Treatment of patients with oligodontia generally requires a multidisciplinary approach. Some patients may require pre-restorative orthodontics. Restoration with a removable partial denture, a conventional fixed partial denture, an implant-retained prosthesis, and adhesive restorative techniques, or a combination of these therapies, are the treatment options. A number of factors must be taken into account for treatment planning. The age of the patient is the most important factor during treatment planning. Other conditions that must be evaluated include the number and condition of the retained teeth, the number of missing teeth, the presence of carious teeth, the condition of supporting tissues, occlusion, and the interocclusal rest space.[15]

However in the present cases, no active treatment was instituted at the first visit, as the retained deciduous teeth did not show any significant root resorption. In the future, after the shedding of primary teeth or the attrition of primary teeth leading to reduced vertical dimension of the face, treatment options for this case would be over dentures with support from erupted permanent maxillary first molars and mandibular second molars or implant-supported prosthesis depending upon the patient's needs and expectation.

Summary

Twins showed the similarity in pattern of hypodontia supporting the strong genetic link. This fact should lead the practitioner to examine other family members. Proper treatment at the appropriate time should be instituted to avoid psychological, esthetic, and functional consequences in patients with oligodontia.

Clinical Significance

Strong genetic link associated with oligodontia help the dentist to know the possibility of its occurrence in other family members and in future generations.

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