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

Rohit B. Gadda, BDS; Keerthilatha M. Pai BDS, MDS; Amar A. Sholapurkar BDS, MDS, FAGE

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.

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. Other frequently used terms are oligodontia, partial anodontia, and anodontia. Oligodontia is defined as agenesis of six or more teeth excluding third molars. Anodontia, the total
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
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).

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.
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. 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.

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

The most distinguishing feature of MSX1-associated oligodontia is the frequent (75%) absence of maxillary first bicuspid, while the most distinguishing feature of PAX9-associated oligodontia is the frequent (>80%) absence of the maxillary and mandibular second molars. 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. Hence cases of oligodontia should be screened and followed for colorectal neoplasia.

Hattab 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. 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.

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

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.

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.

References


About the Authors

Rohit B. Gadda, BDS

Dr. Gadda is a postgraduate student in Department of Oral Medicine and Radiology at the Manipal College of Dental Sciences in Manipal, Karnataka, India. He has authored a few international and national publications. His current research interests include recent advances in the diagnosis of oral cancer. He is a life member of the Indian Academy of Oral Medicine and Radiology.

e-mail: rohitgadda@gmail.com

Keerthilatha M. Pai BDS, MDS

Dr. Pai is a professor and head of the Department of Oral Medicine and Radiology at the Manipal College of Dental Sciences in Manipal, Karnataka, India. She has authored more than 50 international and national publications. Her current research interests include recent advances in the diagnosis of oral cancer. She is a life member of the Indian Academy of Oral Medicine and Radiology.

e-mail: omr@manipal.edu

Amar A. Sholapurkar BDS, MDS, FAGE

Dr. Amar A Sholapurkar is an Assistant Professor in the Department of Oral Medicine & Radiology, Manipal College of Dental Sciences, Manipal, Karnataka, India. He currently serves as a reviewer for The Journal of Contemporary Dental Practice, Journal of Applied Oral Sciences, Oral Oncology, Journal of Medicine and Medical Sciences, Indian Journal of Dental Research, and Manipal Odontoscope.

His research interests include non-surgical management of orofacial lesions and recent advances in the diagnosis and management of oral cancer. He received few best paper awards in the national conferences and has authored more than 23 international publications and 10 national publications. He is a life member of the Indian Academy of Oral Medicine and Radiology and Indian Dental Association.

e-mail: dr.amar1979@yahoo.co.in