Non-Syndromic Oligodontia: Report of Two Cases and Literature Review
Vijaykumar Biradar, Surekha Biradar

Abstract
Dental agenesis is the most common developmental anomaly in humans and is frequently associated with several other oral abnormalities. In the literature, some terms are used to describe missing teeth like oligodontia, anodontia and hypodontia. Oligodontia is defined as the developmental absence of six teeth or more, excluding third molars. It can be isolated or as a part of a syndrome such as in ectodermal dysplasia. The present two cases describe agenesis of permanent teeth which are non-familial and with no apparent systemic abnormalities.

Key Words: Agenesis; Anodontia; Oligodontia; Oral Abnormalities; Hypodontia; Developmental Anomaly.

Introduction
Agenesis of the permanent teeth is an anomaly that is frequently seen in humans. The prevalence of agenesis of one or two teeth (excluding the third molars) is from 1.6% to 9.6%. Oligodontia is a severe type of tooth agenesis involving six or more congenitally missing teeth, excluding the third molars. Although oligodontia is a rare congenital disorder, treatment for this abnormality can be challenge. Specific terms are used to describe the nature of tooth agenesis, like Hypodontia; one to six teeth missing (excluding third molars). Oligodontia more than six teeth are missing (excluding third molars) and anodontia meaning complete absence of teeth. Prevalence becomes progressively smaller as the number of missing teeth increases as seen in oligodontia, with an overall prevalence of 0.14%. A higher frequency has been noted in females than males with no difference in the distribution of missing teeth over maxilla / mandible and left and right sides

Oligodontia can occur in association with genetic syndromes, such as ectodermal dysplasia, Incontinentia pigmienti, Down syndrome and Rieger syndrome or as a non-syndromic isolated familial trait, or as a sporadic finding. When oligodontia is associated with a syndrome there may be abnormalities of the skin, nails, eyes, ears and skeleton. Patients suffering from oligodontia may have several Psychological, esthetic and functional problems. The purpose of this article is to report two cases of congenitally missing permanent teeth and thus the diagnosis of hypodontia/Oligodontia should be performed as early as possible in order to prevent aesthetic and functional problems in dentition.

Case report
Case 1
A 21 year-old female visited to the outpatient department along with her parents for seeking treatment for unpleasant smile and replacement of missing teeth. Patient history revealed no significant past medical and dental history. The patient was normal in her facial appearance and did not show any physical or skeletal abnormality. (Fig 1a) The family history did not reveal any multiple missing teeth. Patient was born to non-consanguineous parents. There was no history of any infection or trauma to the anterior region. Patient mother gave a history of presence of milk teeth in the lower anterior region but after their exfoliation, permanent teeth did not erupt. Patient was healthy with no relevant medical and family history. Suspecting the congenital absence of permanent teeth panoramic radiograph was taken which showed presence of 18 teeth. Teeth present were #17, 15, 14, 13, 23, 24, 25, 26, 27, 34, 35, 37, 38, 44, 45, 46, 47, and 48. (Fig 1b & 1c) Queries revealed that missing lower 6 teeth like #31, 32, 33, 41, 42 and 43 were not extracted, were absent since childhood. The other missing teeth like 11, 12, 16, 21, 22 and 36 were extracted due to caries. Third molar tooth buds in maxillary arch were absent in comparison to mandibular arch. Based on clinical and radiological examinations, diagnosis of nonsyndromic oligodontia was made. The condition was explained to the patient and treatment plan was discussed.

©2012 International Journal of Oral and Maxillofacial Pathology. Published by Publishing Division, Celesta Software Private Limited. All Rights Reserved
Case 2
A 19–year old female patient visited to the outpatient department with major complaint of mastication issues that resulted in dietary deficiencies and esthetics. General physical examination revealed no other abnormalities suggestive of any syndromes. (Fig 2a) Intraoral examination revealed absence of teeth #14, 15, 16, 17, 22, 27, 31, 33, 34, 35, 37, 42, 44, 45, 46 and 47 and teeth present were #11, 12, 13, 55, 21, 62, 23, 24, 25, 26, 71, 32, 36, 41 and 43 with underdeveloped maxillary and mandibular ridges and relatively enlarged tongue (Fig 2b). Radiographic examination confirmed the above missing and retained teeth #55, 62 and 71 as well as underdevelopment of alveolar ridges (Fig 2c). There was no other significant past medical and dental history, neither there was any abnormality detected on general examination suggestive of any syndromes Parental history revealed that there were no other cases of oligodontia in her family. From the above findings diagnosis of nonsyndromic oligodontia was made. To improve appearance, mastication and speech, it was decided that removable partial maxillary and mandibular dentures would be appropriate for the patient.

Figure 1: The clinical photographs showing extra-oral (a) and intra-oral profile with missing teeth in both maxillary and mandibular arches (b) along with orthopantomograph confirming the congenitally missing teeth #31, 32, 33, 41, 42 & 43 (c).

Discussion
Tooth agenesis, the congenital absence of one or more permanent teeth, is a common human anomaly. In most populations, the reported prevalence of permanent tooth agenesis, excluding third molars, varies from 2.2 to 10.1%. In the majority of cases, persons are missing only one tooth. The prevalence becomes progressively smaller as the number of missing teeth increases. Agenesis of more than two teeth occurs in approximately 1% of the population. Selective tooth agenesis is divided into 2 types: hypodontia, the agenesis of fewer than 6 teeth, and oligodontia, the agenesis of six or more permanent teeth. In both cases, the third molars (wisdom teeth) are not included. Oligodontia is a rare anomaly, affecting approximately 0.1 to 0.3% of the population.

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.\(^7\) Oligodontia as well as hypodontia (lack of one or more permanent teeth) are highly heritable conditions associated with mutations in the AXIN2, MSX1, PAX9, EDA, and EDAR genes.\(^8\) The incidence of oligodontia is reported to vary from 0.08 - 0.16%. The pattern of tooth absence is influenced by the gene affected, as well as the type of mutations within the specific gene.\(^9\) 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 virus infections, toxins and radio or chemotherapy may cause missing of permanent teeth. However, most of the cases are caused by genetic factors.\(^5\) Although several potential and verified environmental factors in tooth agenesis have been identified, genetic defects play a major role in the etiology. So far, researchers have identified genetic defects that cause tooth agenesis either as a sole anomaly (isolated or non-syndromic) or as a part of multiple congenital anomalies (syndromic). One gene associated with syndromic tooth agenesis is the EDA gene, which underlies X-linked hypohidrotic ectodermal dysplasia. Non-syndromic tooth agenesis has wide phenotypic heterogeneity and is classified as either sporadic or familial, which can be inherited in an autosomal-dominant, autosomal-recessive, or X-linked mode.\(^6\)

Figure 2: The clinical photographs showing extra-oral (a) and intra-oral profile with missing teeth in mandibular arch (b) along with orthopantomograph confirming the missing permanent and retained deciduous teeth (c).

The present case reported absence of lower anterior permanent teeth and the use of panoramic radiography was recommended, together with clinical examination for the detection or confirmation of dental development and performing the diagnosis of oligodontia. In the reported case, panoramic radiography revealed absence of twelve permanent teeth. Out of which lower six anterior teeth were congenitally missing with no other associated dental anomalies like delayed formation and eruption of teeth, reduction in tooth size and form, ectopic eruption of teeth, rotation of teeth except for enamel hypoplasia. Because of its variable genetic etiology, the presentation and subsequent clinical effect on the dentofacial structures of patients diagnosed with oligodontia may vary greatly.\(^5\) Congenital missing teeth may create dental and facial disfigurement, which can lead to social withdrawal, especially in adolescent years.\(^2\) Prosthodontic treatment of oligodontia patients is, therefore, important for functional, esthetic
and psychological reasons. Treatment of such patients requires a fully integrated interdisciplinary approach of orthodontists, oral and maxillofacial surgeons and prosthodontist. 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 interocclusal space. In the present case, impression of mandibular arch was made and the patient was asked to come back in the next visit. But the patient was lost for follow up. Such an approach becomes a major hindrance to successful treatment of such cases.

In case 2, early prosthetic treatment led to significant improvements in appearance, speech, and masticatory function. During the first month following the initiation of prosthetic treatment, it was difficult for the patient to adapt to the removable partial dentures; however, she was accustomed to using the dentures and was able to eat adequately within a few months. Although dentures are poor alternatives to healthy dentition, they create conditions for the maintenance of a normal, satisfactory diet for the individual.

Conclusion

Oligodontia is frequent finding in many syndromes, but in this case it was not associated with any syndrome which was a rare finding. So as an oral physician if we come across any case having multiple congenital missing teeth it may not be always associated with multiple other abnormalities as seen in syndromes. Patients of oligodontia generally require a multidisciplinary approach. Thus, the cases should be evaluated carefully by clinicians and early diagnosis and treatment planning should be made for appropriate treatment modalities and to minimize the complication of these dental anomalies.

Acknowledgement

We would like to thank all the staff members of the department of oral pathology for their constant support and encouragement.

Author Affiliations

1. Dr Vijaykumar Biradar, Senior Lecturer, Department of Oral and Maxillofacial Pathology, 2. Dr Surekha Biradar, Lecturer, Department of Oral Medicine and radiology, Maharashtra University of Health Sciences, MIDSR Dental College, Hospital and Research Centre, Latur, Maharashtra, India.

References


Corresponding Author

Dr Vijaykumar Biradar
Senior Lecturer, Department of Oral Pathology, MIDSR Dental College, Hospital and Research Centre, Latur - 413512, Maharashtra, India.
Email: dvijay06_biradar@yahoo.com
Ph: +919890459777

Source of Support: Nil, Conflict of Interest: None Declared.