

CASE REPORT



Non-syndromic oligodontia: A case report

Tejavathi Nagaraj, Soniya Kongbrailatpam, Yasir Shafeeq Mohiyuddin, Ijum Doye

Department of Oral Medicine and Radiology, Sri Rajiv Gandhi College of Dental Science and Hospital, Cholanagar, Bengaluru, Karnataka, India

Correspondence:

Dr. Soniya Kongbrailatpam, Department of Oral Medicine and Radiology, Sri Rajiv Gandhi College of Dental Science and Hospital, Cholanagar, Bengaluru, Karnataka.
E-mail: soniya.kongbrailatpam00@gmail.com

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Abstract

Oligodontia is defined as the developmental absence of six teeth or more, excluding third molars. Anodontia is defined as complete absence of teeth, and hypodontia is missing one to five teeth. The consequences of missing teeth include abnormal occlusion or altered facial appearance, which can cause psychological distress in some patients. Orthopantomograph and lateral skull radiograph are useful to view the development of primary and permanent tooth buds and jawbones. A multidisciplinary approach is required to complete oral rehabilitation, which includes orthodontic treatment, surgical intervention, prosthodontic rehabilitation, and psychological counseling.

Keywords: Oligodontia, panoramic radiograph, primary and permanent dentition**Introduction**

Oligodontia can be classified as syndromic or non-syndromic. Non-syndromic oligodontia is a developmental dental anomaly without the involvement of other organs.^[1,2] The exact etiology of oligodontia is unknown. Dental agenesis is a common developmental anomaly that affects approximately 20% of the population and results in a reduction of the number of teeth present in the oral cavity.^[3] It is classified based on the number of missing permanent teeth, excluding the third molars. In most populations, Polder *et al.* 2004 reported prevalence of permanent tooth agenesis, excluding third molars, varies from 2.2 to 10.1%.^[4] The reported incidence of teeth other than third molars being missing varies from 1.6% to 9.6%.^[3] The most commonly missing permanent teeth are the third molars (9–37%), followed by mandibular second premolars (<3%), maxillary lateral incisors (<2%), and maxillary second premolars and mandibular incisors (<1%).^[3] The present case report shows missing permanent teeth and retained deciduous teeth with non-familial and no apparent abnormalities.

Case Report

A 21-year-old male patient reported to the department of oral medicine and radiology with a chief complaint of missing teeth in the lower front teeth region since birth and wanted to get them fixed. There was no history of extraction of any tooth; family history revealed non-consanguineous marriage with normal delivery.

On extraoral examination, the patient's skin and hair were found to be normal in texture and appearance [Figure 1].

Intraoral examination reveals 7 missing permanent teeth, that is, 15, 25, 31, 35, 41, 44, and 45. Teeth present are 11, 12, 13, 14, 16, 17, 21, 22, 23, 24, 26, 27, 32, 33, 34, 36, 37, 42, 43, 46, and 47. Retained deciduous teeth are 55, 75, 84, and 85 [Figures 2 and 3].

Orthopantomograph reveals teeth present 11, 12, 13, 14, 16, 17, 21, 22, 23, 24, 26, 27, 32, 33, 34, 36, 37, 42, 43, 46, and 47. Retained deciduous teeth are 55, 75, 84, and 85 [Figure 4]. There are no impacted teeth present. Based on the above clinical findings, a provisional diagnosis of non-syndromic oligodontia was made.

To rule out other etiologies, complete blood investigations, serum Ca, alkaline phosphate, and thyroid function tests were advised. The findings of these investigations were normal. Based on the reports, non-syndromic oligodontia was given as the final diagnosis. The patient was referred to the department of oral and maxillofacial surgery for further management.

Discussion

Oligodontia is defined as the developmental absence of six teeth or more, excluding third molars. Oligodontia can be classified as syndromic or non-syndromic. Non-syndromic oligodontia is a developmental dental anomaly without the involvement of other organs. Non-syndromic oligodontia has been shown to be inherited as an autosomal dominant trait. 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, Ectodactyly ectodermal dysplasia-clefting syndrome, cleft lip palate ectodermal dysplasia syndrome,



Figure 1: Extraoral picture



Figure 2: Intraoral examination of maxillary teeth

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.^[5] When oligodontia is associated with a syndrome, there may be abnormalities of the skin, nails, eyes, ears, and skeleton.^[5] Damage to dental lamina before tooth formation can result in hypodontia. Hypodontia is the lack of development of one or more teeth. Causes of hypodontia include genetic (autosomal dominant, recessive, or sex-linked patterns), trauma, endocrine disturbances, infection, radiation, and chemotherapeutic medications. It usually affects permanent third molars, second premolars, and lateral incisors in that order. Associated microdontia may be observed. Hypodontia may cause the abnormal spacing of teeth, delayed tooth formation, delayed deciduous tooth exfoliation, and late permanent tooth eruption. Prosthetic replacement of teeth may be needed. Etiopathogenesis shows that the exact etiology of oligodontia is unknown. Heredity is one of the main etiological factors.^[6] Other factors include environmental factors such



Figure 3: Intraoral examination of mandibular teeth



Figure 4: Orthopantomograph

as viral infections and irradiation, or chemotherapy can affect the formation of permanent teeth.^[6] Abnormal gene function may also disrupt specific signaling pathways involved in tooth development, resulting not only in abnormal tooth number but also in abnormal tooth size and or shape. 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.

The genes and molecular pathways involved in tooth agenesis are Wnt/b-Catenin/LEF1, MSX1, MSX2, SHH p63 Pitx2, Runx 2/Cbfa1, and PAX9 genes.^[7] The MSX1 gene is located in chromosome number 4 which is involved in several epithelium-mesenchymal processes of embryogenesis.^[6] This gene is commonly expressed in the mesenchyme of the tooth buds, during its early stages.^[6] The PAX9 gene is located on the chromosome 14 and belongs to a family of genes which are involved in transcription factors which act during the initiation of the embryo's development.^[6] Expression of PAX9 gene is required for dental mesenchyme to condense around the epithelium of developing tooth bud. Protein products of the PAX9 and MSX1 genes play a major role in the maintenance and expressions of mesenchymal bone morphogenetic protein 4, which initiates the morphogenesis of the dental organ.^[6] Defects in the PAX9 and mutations in the MSX1 gene have been identified in humans; thus, once there is an expression defect of these genes, there might be an arrested of tooth bud development, leading to lack of development of tooth bud.^[8]

Conclusion

Oligodontia cases should be reported so as to enhance the understanding of this rare entity. It can lead to functional, esthetic, and social loss to a patient; hence, a prompt treatment can lead to improved quality of life.

Declaration of Patient Consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient had given his consent for his images and other clinical information to be reported in the journal. The patients understood that their names and initials will not be published, and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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