Case Report

Non-Syndromic Oligodontia Of Primary And Permanent Dentition: A Case Report

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ABSTRACT

Oligodontia is generally defined as the congenital absence of six or more permanent teeth, excluding the third molars. It is a rare dental anomaly with a prevalence of 0.3% in permanent teeth and much less frequency in the primary dentition. It is usually a part of a syndrome and seldom occurs as an isolated entity. Genes responsible for non syndromic oligodontia are found to be MSX1 and PAX9 genes. A female patient aged 12 years reported with 6 deciduous and 6 permanent teeth missing excluding third molars. Early recognition and adequate treatment is paramount for the proper function and esthetics of the patient.

Key words: Anodontia, hypodontia, oligodontia, dental agenesis.

Introduction

True anodontia or congenital absence of teeth are of two types, total and partial. Total anodontia, in which all teeth are missing may involve both the deciduous and the permanent dentition. This is a rare condition; when it occurs it is frequently associated with a more generalized disturbance, hereditary ectodermal dysplasia.[1] Induced or false anodontia occurs as a result of extraction of all teeth while the term pseudoanodontia is sometimes applied to multiple unerupted teeth.[1]

True partial anodontia (hypodontia or oligodontia involves one or more teeth and is a rather common condition. Although any tooth may be congenitally missing, there is a tendency for certain teeth to be missing more frequently than others.

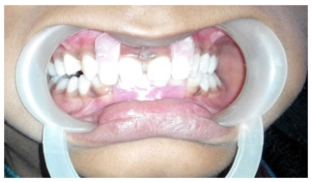
Studies on the frequency of missing third molars have shown this tooth to be congenitally absent in as many as 35% of all subjects examined with a frequency absence of all four third molars in the same person. Other studies have shown that the maxillary lateral incisors and maxillary or mandibular second pre-molars are commonly missing, often bilaterally. In severe partial anodontia, the bilateral absence of corresponding teeth may be striking. An outstanding review of this subject has been reported by Graber, who showed that overall frequency of patients with congenitally missing teeth (excluding third molars) has ranged from 1.6 to 9.6 per cent in various series of studies in different countries.[2] Congenitally missing deciduous teeth are uncommon but, when occurring usually involve the maxillary lateral incisor. Mandibular lateral incisors and mandibular cuspids may also be missing, according to the study of Granath. Their studies also show a close correlation between congenitally missing deciduous teeth and their permanent successors, suggesting at least in some instances a genetic factor.[3]

Oligodontia is the severe form of hypodontia in which there is congenital absence of six or more teeth (excluding third molars).[4]

In most populations, the reported prevalence of permanent tooth agenesis, excluding third molars, varies from 2.2-10.1%. [Polder, et al,. 2004].[5]As the number of missing teeth increases the prevalence becomes progressively smaller. The incidence of oligodontia is reported to vary from 0.08 to 0.16% .[6] Oligodontia is often associated with specific syndromes affecting several body organs and systems. Oligodontia can be classified as non-syndromic (isolated) and syndromic.

Isolated form can be familial or sporadic in nature. There are more than 49 syndromes associated with congenital absence of teeth. When oligodontia is a part of syndrome, concomitant abnormalities of skin, hair, nail, eyes or skeleton are present in the patient. The pattern of tooth absence is influenced by the gene affected, as well as the type of mutations with in the specific gene[7].

Oligodontia can occur in association with various genetic syndromes, such as ectodermal dysplasia, incontinentia pigmentia, Down syndrome, Rieger syndrome, Wolf–Hirschhorn syndrome, Van der Woude syndrome, Ectodactyly-ectodermal dysplasia-clefting syndrome, cleft lip palate ectodermal syndrome, oral facial digital syndrome type I, Witkop's tooth and nail syndrome, Fried syndrome, hair-nail-skin-teeth dysplasias, Hirschhorn syndrome, hemifacial microsomia,



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Discussion

In this report, the patient presented with the congenital absence of 6 permanent and 6 primary anterior teeth. Studies on the prevalence of Oligodontia have shown that more than 80% of the population will present with one or two congenitally missing teeth while less than 1% will present with six or more teeth missing.[6] There are very few cases reported in literature in which primary teeth are congenitally missing. Studies have shown that the most commonly missing teeth are permanent premolars and maxillary lateral incisors but in this case all permanent premolars were present.[7]Another significant finding was that the pattern of missing teeth was symmetrical.

Entire management of all these dental problems in a patient afflicted with oligodontia is through an multidisciplinary path i.e by involvement of team comprising of a pediatric dentist, orthodontist, prosthodontist and an oral & maxillofacial surgeon[,9,10,11]

Prosthodontic treatment of Oligodontia can include removable or fixed partial dentures or implant supported prosthesis.[12]

Congenital absence of teeth can create dental and facial disfigurement, which can lead to social withdrawal.[13]Normal psychological development of the child with oligodontia is also a major matter. Therefore, the removable partial denture fabricated for the patient in this case would help her to improve her peer group relationship, guide and prevent her from developing abnormal speech and tongue habits. Early rehabilitation with interim prosthesis will help the child interact better with his peers and society.Prevalent methods of treatment employed include, prerestorative orthodontics which is frequently required to move teeth to a favourable position, restoration with removable or fixed partial dentures and restoration with implant-supported prosthesis.[14,15]

Final treatment with osseointegrated implants and supplemental bone augmentation procedures has shown successful treatment outcomes in such cases. Implants can be placed once the skeletal growth of the jaws has been completed because implants inserted into pediatric patients do not follow the regular growth process of the craniofacial skeleton and are known to behave similar to ankylosed teeth, resulting in both functional and esthetic disadvantages (OpHeji et al, 2003).[16] They can interfere with the position and eruption of adjacent tooth germs, thus resulting in potential severe trauma to the patient.When placed in alignment with adjacent teeth, the implants did not participate in growth processes, resulting in an infra-occlusionand multidimensional dislocation when compared with the developing teeth (Sennerby et al, 1993).[17]Aditionally adjacent tooth germs exhibited morphologic changes and disorders of eruption (Thilander et al, 1992)[18].

Although the etiology of single missing teeth is unknown, a familial tendency for this defect is present in many instances. Graber in reviewing congenital absence of teeth, reported the accumulating evidence that it is actually the result of one or more point mutations in a closely linked polygenic system, most often transmitted in an autosomal dominant pattern with incomplete penetrance and variable expressivity.Heredity being major factor in Oligodontia as well as hypodontia (lack of one or more permanent teeth) are highly associated with mutations in the AXIN2, MSX1, PAX9, EDA, and EDAR genes.[19] The genes and molecular pathway involved in tooth agenesis are Wnt/b-Catenin/LEF 1 MSX 1 MSX 2 SHH p63 Pitx 2 Runx 2/Cbfa 1.[20]

Conclusion

Oligodontia has a substantial influence on speech, mastication and psychological maturation of the child. In such a case,early and accurate diagnosis is necessary as well as the careful planning of treatment for the best outcome.

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and recessive incisor hypodontia.[8]

Case Report

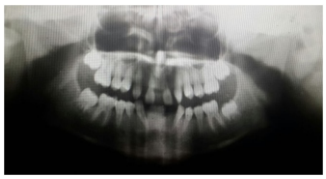
A female patient aged 12 years reported to the Department of Paedodontics and Preventive Dentistry, Rama Dental College Hospital and Research Centre with a chief complaint of missing teeth in both upper and lower front teeth region of the jaw.Detailed history from the patient and her parents revealed that the patient is teased at school by other students because of her permanently missing teeth and has a poor peer group relationship.

Medical history was not significant. Past dental history revealed that Patient has also undergone ortho treatment which she discontinued after sometime. Extra-oral examination revealed no skeletal abnormality and facial asymmetry. Intra oral examination revealed a large midline diastema and mesially converging maxillary central incisors.

Permanent maxillary lateral incisors and all four mandibular incisors were not present clinically. Periodontal health of the patient was satisfactory. Parents of the patient confirmed that missing teeth were not due to extraction or trauma and primary predecessors of these teeth i.e. 52, 62, 71, 72, 81 and 82 never erupted.

Radiological assessment was performed which revealed that 12,22,31,32,41,42are congenitally absent along with the permanent buds of third molars.

Clinical and radiological assessment of maxillary and mandibular alveolar bone revealed that mandibular alveolar bone is very thin and deficient.



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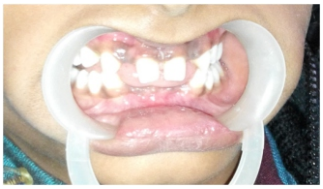

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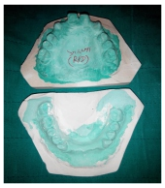

Study Models

Thorough physical evaluation was performed with particular attention to the skin, hair, nails, eyes and ears of the patient all of which appeared normal.

There was no skeletal abnormality. Parents of the patient reported no sweating abnormality. Family history was non-significant.

A total number of six permanent teeth (12, 22, 31, 32, 41and 42) and six primary teeth (52, 62, 71, 72, 81 and 82) were missing congenitally.

A diagnosis of a case of 'non-syndromic oligodontia of primary and permanent dentition'was made.

After a thorough evaluation and study of the patient's case, provision of an interim prosthesis for both the mandibular and maxillary arch was planned as the first line of treatment.

A consultation from Orthodontics department was made where an initial orthodontic evaluation was thoroughly performed. It was decided that patient will have to undergo fixed orthodontic wire therapy for the closure of the wide midline diastema and to create space for the permanent prosthesis. Osseointegrated implants were selected as the final prosthesis for all the congenitally missing teeth. Implants shall be placed once the skeletal growth of the jaws has been completed.

Patient and both the parents of the patient wanted an esthetically acceptable interim prosthesis for the upper and lower arch for rehabilitation for 3-4 months before undergoing ortho treatment since the patient was not welloff financially and wanted time to finance the ortho treatment. Therefore, for better function and esthetics a removable acrylic partial denture was fabricated to function as an interim prosthesis for both upper and lower arch. Pre-treatment occlusal study of the upper and lower diagnostic cast was performed. Upper and lower arch impressions were taken with alginate. Dental stone is poured up to make the working cast. After curing, finishing and polishing, removable partial dentures were delivered. Occlusion checked for any premature contacts.

Post treatment instructions were given to the patient. The patient appeared happy on receiving the dentures and her parents showed satisfaction towards the treatment. Regular 3 months follow up visits were scheduled for complete management of the condition.

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