Multidisciplinary Treatment Approach of A Rare Case of Familial Non Syndromic Oligodontia - A Case Report

Dr. Indrani Das

Department of Prosthodontics , Regional Dental College,Guwahati,Assam

Access Online

Abstract

Oligodontia is the congenital agenesis of 6 or more permanent teeth,excluding third molars.The prevalence of oligodontia has been reported as 0.3% and affects females more often than males,with a gender ratio of 3:2.Although oligodontia can occur over with 60 different syndromes,these anomalies can occur without any syndrome or systemic disease.All mutations of PAX9 identified to date have been associated with nonsyndromic form.This case report describes a multidisciplinary treatment approach toward a female patient 16 years of age presenting with familial nonsyndromic oligodontia having paternal family trait and absence of 28 permanent teeth.The treatment plan considered for the patient included extraction of deciduous teeth with resorbed roots and poor prognosis. Remaining deciduous teeth which were affected by secondary caries were restored by endodontic therapy.Finally rehabilitation was done by removable partial denture for maxillary arch and overdenture for mandibular arch.It is emphasized that conventional prosthetic treatment can lead to a satisfactory result.

Keywords: Oligodontia, Overdenture, Deciduous teeth, Agenesis

Introduction

Agenesis of teeth is one of the most common of human developmental anomalies. Oligodontia is the congenital agenesis of 6 or more permanent teeth, excluding third molars.Oligodontia has been classified as isolated, familial or nonsyndromic (which is more common) and syndromic hypodontia (as ectodemal dysplasia, Rieger's syndrome, Witkop syndrome , oro-facial-digital syndrome, or incontinentia pigmenti). All mutations of PAX9 identified to date have been associated with nonsyndromic form of tooth agenesis. Tooth agenesis (MIM# 106600), the congenital absence of one or more permanent teeth, is a common human anomaly (Pemberton, et al.[1], 2005). In most populations, the reported prevalence of permanent tooth agenesis, excluding third molars, varies from 2.2-10.1% (Polder, et al.[2], 2004). In the majority of cases, persons are missing only one tooth (Daugaard-Jensen, et al.[3], 1997). 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 (Polder, et al.[2], 2004). Oligodontia (MIM# 604625) is a rare anomaly, affecting approximately 0.1 to 0.3 % of the population (Polder et al.[2], 2004).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 (Burzynski and Escobar[4],1983). Dominant mutations in MSX1 (MIM# 142983), PAX9 (MIM# 167416), and AXIN2 (MIM# 604025) have been found in families with non-syndromic tooth agenesis (; Stockton et al., 2000, Lammi et al.[5], 2004) . However, mutations in these genes were detected in only a few affected individuals, suggesting that there may be other unidentified genetic defects responsible for this disease (Scarel et al.[6], 2000; Frazier-Bowers et al.[7], 2002). The purpose of this article is to report a multidisciplinary treatment approach toward a 16 year old female patient with a rare case of congenitally missing permanent teeth.

Pre-treatment evaluation of the patient:

1)Extra-oral examination of the patient revealed facial asymmetry and decrease in lower facial height and flat profile [Fig 1]. The nasolabial angle was within normal limits, the lower lip full and slightly protruded relative to the upper lip at closure, a deep mentolabial sulcus was present and increased activity of the mentalis muscle was visible in the skin overlying the chin upon lip closure.

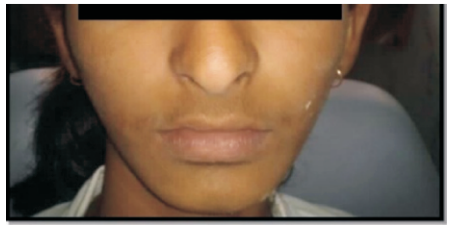
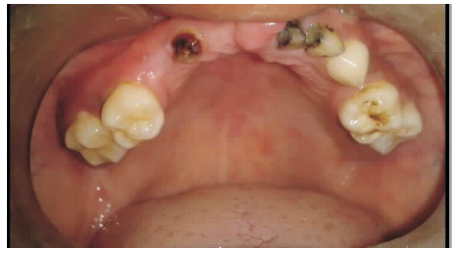


Fig 1 Extraoral view

2)Intra-oral examination revealed that all permanent teeth were missing since childhood and not due to extraction. Teeth present in maxilla were deciduous 45,46,55,56,53,and root stumps of 42,51,52 . Teeth present in mandible were 73, 74,81,82,83 [Fig 2(a&b)]



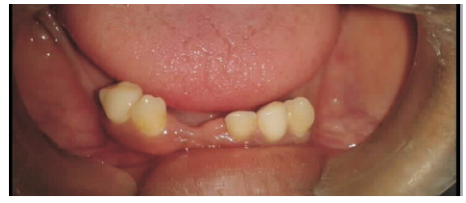


Fig 2 (a&b) Intraoral view

Radiographic examination revealed all teeth in various stages of caries and root resorption. Orthopantomogram showed missing permanent maxillary and mandibular teeth. [Fig 3(a)]

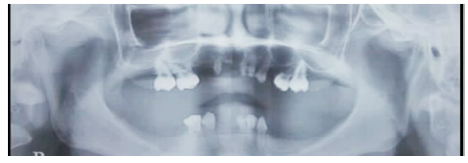
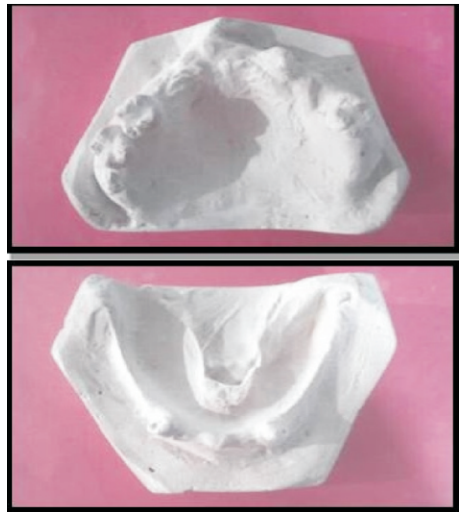


Fig 3 (a) OPGFig 3



(b) Study Casts

Genetic counseling

The patient is the second child of 2 from a non-consanguineous marriage. Patient's father and grandfather had oligodontia. The patient's neuromotor development appeared normal and there was no specific finding on physical examination to suggest any syndrome. Ophthalmic examination and skeletal X-ray indicated normal findings.

Case pedigree analysis revealed presence of oligodontia in previous generations and absence of oligodontia in sibling suggests X-linked dominant inheritance.

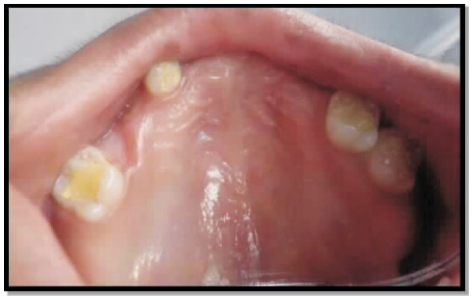
Complete set of investigations were done which included routine examination of blood including serum calcium, alkaline phosphate, TSH, T3, T4 , findings of which were normal. During physical examination, hairs were not thin and sparse, nails were not brittle and no difficulty in perspiration was seen which ruled out ectodemal dysplsia; on ocular examination, no signs of glaucoma was seen ruling out Rieger syndrome and lastly Van Der Woude syndrome was left out as there was no cleft palate or any mucosal cysts in lower lip. Hand wrist radiographic examination was normal. Finally based on above findings Non syndromic Oligodontia as final diagnosis was justified.

The treatment plan considered for the patient was divided into four phases:

PHASE I Oral surgical
PHASE II Periodontic
PHASE III Endodontic phase
PHASE IV Prosthetic rehabilitation

Phase I-Oral surgical:

This phase included extraction of deciduous teeth with resorbed roots and poor prognosis. [Fig 4 (a&b)]



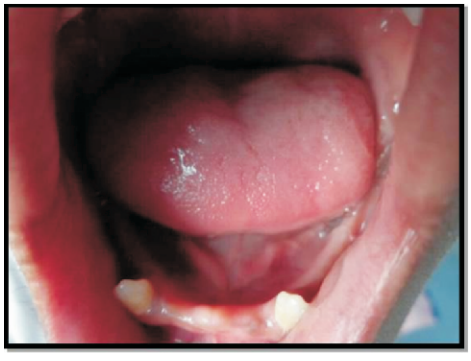
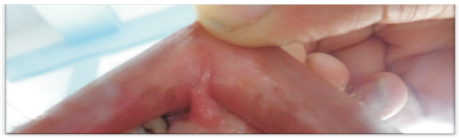


Fig 4 (a&b) Extraction of teeth with poor prognosis

Phase II - Periodontic [Fig 5(a&b)] Frenectomy was performed in relation to maxillary labial frenum to correct high frenal attachment.



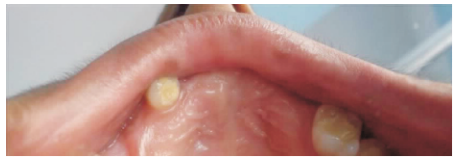
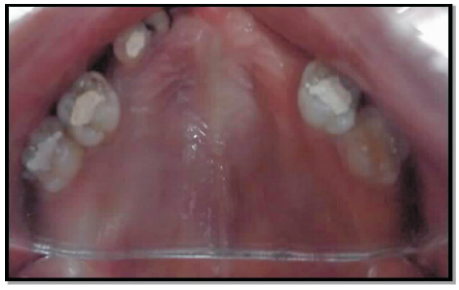


Fig 5 (a&b) Frenectomy performed in relation to maxillary labial frenum

Phase II-Endodontic phase:

After healing of extraction sites, remaining deciduous teeth which were affected by secondary caries were restored by endodontic therapy. An endodontically treated maxillary left 2nd molar tooth had surgical crown lengthening on the palatal side due to insufficient crown height. [Fig 6 (a&b)]



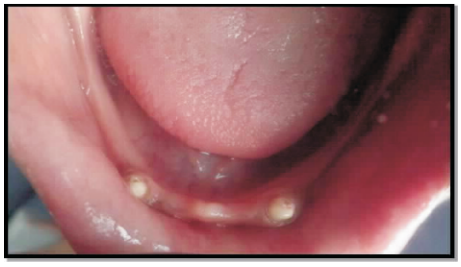
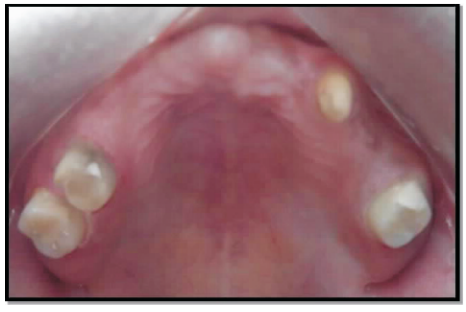


Fig 6 (a&b) Endodontic treatment of carious affected teeth

Phase IV—Prosthetic phase:

After 3 weeks of gingival healing maxillary right canine, and molars were prepared to receive crowns, and mandibular canines were prepared to receive overdenture copings and serve as overdenture abutments [Fig 7 (a&b)]. Finally removable partial denture for maxillary arch and overdenture for mandibular arch was being considered for aesthetic and functional rehabilitation of the patient.



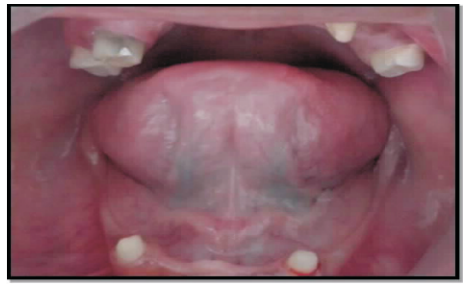
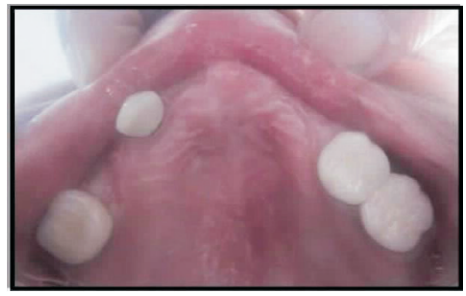


Fig 7 (a&b) Tooth preparation of maxillary and mandibular teeth

Porcelain fused to metal restorations were fabricated for maxillary teeth and cast metal copings were fabricated for mandibular teeth and cemented using Glass ionomer cement (type I) [Fig 8 (a&b)]



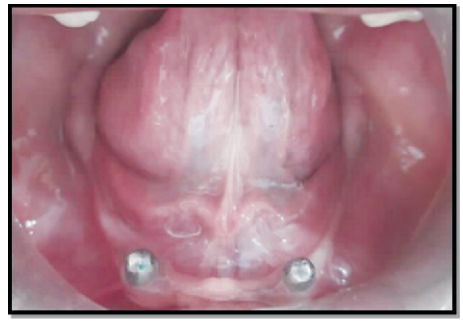


Fig 8 (a&b) Cementation of crowns to maxillary teeth and cast metal copings on mandibular overdenture abutments

Maxillary impression was made by dual impression technique. Border moulding was done using a single step putty technique and final impression was taken with PVS (light body consistency-aquasil) [Fig 9 (a&b)].

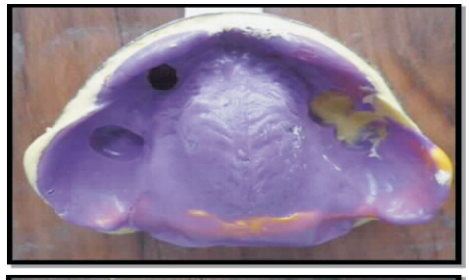
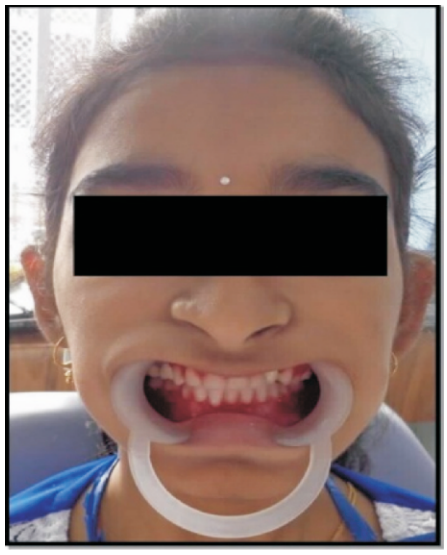


Fig 9 (a&b) Final impressions

For mandibular arch border moulding and final impression were done. The impressions were poured using Type II dental stone. Routine procedures were followed for construction of prostheses [Fig 10 (a)]. Processed dentures were finished, polished and post insertion instructions were given to the patient and attendants [Fig 10 (b)]. The patient reported that her self-esteem and quality of life have improved.



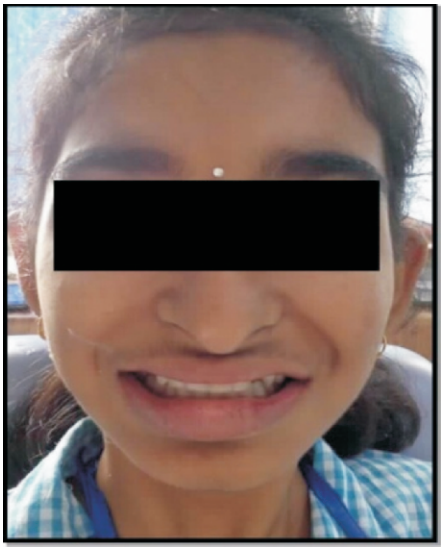


Fig 10 (a&b) Try in and Post insertion view

Discussion

Factors which play a significant role in selecting and planning treatment of oligodontia are:

- Patient age
- number and condition of present teeth,
- number of missing teeth, presence of carious teeth,
- condition of supporting tissues,
- occlusion and inter-occlusal rest space

In the present patient, the persistent deciduous teeth were retained by endodontic therapy to take advantage of their prevention of ridge resorption, In the present case, for biomechanical purposes, the canines in mandibular arch had to be used as abutment teeth under overdenture. Management of oligodontia patients with dental implants is difficult, because the congenital absence of teeth can result in lack of sufficient alveolar ridge for successful implant placement. Nevertheless, oral rehabilitation of oligodontia patients via dental implants is regarded as an efficient and successful treatment.Considering the prosthetic rehabilitation of all missing permanent teeth, the present patient selected conventional tooth tissue supported prosthesis over implant-supported restorations because implant-supported restorations would require bone grafting and secondary surgical procedure for placement of implants to replace the missing teeth and to restore occlusion.

Conclusion

Treatment planning for oligodontia should be individualized for each patient. Prosthetic treatment is rendered in a stage wise manner initiating with removable prostheses and continued till full growth is attained. Treatment protocol during this stage includes periodic follow up for adjustment and remaking of prostheses during rapid growth period. Once full growth is achieved definitive treatment in form of implant supported fixed and/ or removable partial denture may be provided.It is emphasized that conventional prosthetic treatment can lead to a satisfactory result. Every case of oligodontia should be reported so as to enhance the understanding of this rare entity.

References

1. Pemberton TJ, Das P, Patel PI. Hypodontia: genetics and future perspectives. Braz J Oral Sci. 2005; 4:695-706
2. Polder BJ, Van't Hof MA, Van der Linden FP, Kuijpers-Jagtman AM. A meta-analysis of the prevalence of dental agenesis of permanent teeth. Community Dent Oral Epidemiol. Jun 2004; 32(3):217-26.
3. Jensen J, Nodal M, Kjaer I. Pattern of agenesis in the primary dentition: a radiographic study of 193 cases. Daugaard. Int J Paediatr Dent. Mar 1997; 7(1):3-7.
4. Burzynski NJ, Escobar VH. Classification and genetics of numeric anomalies of dentition Birth Defects. Orig Artic Ser. 1983; 19(1):95-106.
5. Vastardis H, Karimbux N, Guthua SW, Seidman JG, Seidman CE. A human MSX1 homeodomain missense mutation causes selective tooth agenesis. Nat Genet. 1996; 13(4):417-21.
6. Stockton DW, Das P, Goldenberg M, D'Souza RN, Patel PI. Mutation of PAX9 is associated with oligodontia. Nat Genet. 2000; 24(1):18-19.
7. Lammi L, Arte S, Somer M, Jarvinen H, Lahermo P, Thesleff I, Pirinen S, Nieminen P. Mutations in AXIN2 cause familial tooth agenesis and predispose to colorectal cancer. Am J Hum Genet. 2004; 74(5):1043-50.
8. Scarel RM, Trevilatto PC, Di Hipólito O Jr, Camargo LE, Line SR. Absence of mutations in the homeodomain of the MSX1 gene in patients with hypodontia. Am J Med Genet. 2000; 92(5):346-9.
9. Frazier-Bowers SA, Scott MR, Cavender A, Mensah J, D'Souza RN. Mutational analysis of families affected with molar oligodontia. Connect Tissue Res. 2002; 43(2-3):296-300.

Figures Legend:

Fig No	*Description*
FIG 1	Extraoral view
FIG 2(a) & 2(b)	Intraoral view
FIG 3(a) & 3(b)	OPG & Study Casts
FIG 4(a) & 4(b)	Extraction of teeth with poor prognosis
FIG 5(a) & 5(b)	Frenectomy performed in relation to maxillary labial frenum
FIG 6(a) & 6(b)	Endodontic treatment of carious affected teeth
FIG 7(a) & 7(b)	Tooth preparation of maxillary and mandibular teeth
FIG 8(a) & 8(b)	Cementation of crowns to maxillary teeth and cast metal copings on mandibular overdenture abutments
FIG 9(a) & 9(b)	Final impressions
FIG 10(a) & 10(b)	**Try in and Post insertion facial view**