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CASE REPORT

Non syndromic congenital agenesis of multiple permanent teeth: Case series and recent literature review

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Introduction: Hypodontia refers to the congenital absence of less than six teeth. This absence may be unilateral or bilateral. Though the congenital agenesis of bilateral mandibular/ maxillary incisors has often been reported in literature, however, the congenital absence of bilateral mandibular and maxillary incisors, as well as the mandibular second molar- giving rise to a total of six missing permanent teeth in an apparently healthy individual has not been reported earlier. **Case presentation**: This case series presents two cases of a 10- and 11-year-old children with presence of retained deciduous anterior in both arches with absence of permanent successors and also aims to review the literature regarding etiology, clinical implications and management in such cases. **Conclusions**: It is essential that practitioners monitor the developing dentition with establishment of a proper review schedule. Non-eruption of the permanent tooth more than one year later than expected, or even after six months following the emergence of the contralateral tooth, warrants a high degree of suspicion. A multidisciplinary team, including pediatric, restorative and orthodontic specialists, is advised. In addition, prior to formulation of any treatment plan, due consideration to the general issues such as the patient's systemic and oral health, motivation and expectations should be given.

Keywords: congenital agenesis, permanent teeth, hypodontia

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Introduction

Primary teeth may be retained for a variety of reasons. The congenital absence of permanent successors, or dental agenesis, is one of the most common causes for the retention of deciduous predecessors [1]. This is encountered relatively frequently with a prevalence of 2.5-6.9% and a female predilection in different races [2]. Agenesis can be classified as hypodontia where there is absence of one to five teeth (except third molars), oligodontia when six or more teeth are missing and anodontia when there is complete absence of permanent teeth. Oligodontia is a relatively rare condition, affecting only about 0.1 to 1.2% of the population [3].

Some teeth are more prone to developmental agenesis as compared to others. Mandibular second premolars are known to be most frequently missing (2.9-3.2%), followed by maxillary lateral incisors (1.6-1.8%), maxillary second premolars (1.4-1.6%) and mandibular incisors (0.2-0.4%), while the absence of other teeth is relatively rare.[2] Clinicians claim that the prevalence of congenital agenesis may be increasing, perhaps due to the evolutionary changes and as a result of more advanced screening methods in diagnosis [4,5].

The etiology of dental agenesis is best described by a multifactorial model.[6] Although predominantly a heritable phenomenon with an autosomal dominant pattern, the severity of expression may be affected by environmental factors such as localized infection of the jaw, endocrinal disturbances, systemic infection, trauma and drugs like thalidomide or chemotherapeutic agent [5].

Oligodontia may also be associated with a particular syndrome like Down's syndrome, anhidrotic ectodermal dysplasia, incontinentia pigmenti, Pierre Robin syndrome, and Ehler- Danlos syndrome [6].

Although in routine dental practice, these anomalies account for a relatively low frequency of occurrence, as compared to the more common oral pathologies such as dental caries and periodontal diseases, nevertheless, they can pose a problem during treatment planning. Clinical management is usually complicated with complaints of occlusal, aesthetic and functional problems, in addition to a possible disposition for other oral diseases [7].

Therefore, the aim of the present article is to report 2 cases of oligodontia due to congenital agenesis of permanent teeth in non-syndromic, apparently healthy children and to review the literature in relation to its etiology, clinical implications and management.

Case 1

A 11-year-old male patient reported to our department with complaint of food lodgment in the lower left back tooth for the past 1 month. There was no history of associated pain/ sensitivity. The patient's medical history was unremarkable. On clinical examination, there was a deep carious lesion on 74 that necessitated extraction. However,

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apart from the region of chief complaint, it was unusual and interesting to note that, in the maxillary arch, the permanent central incisors had erupted with presence of bilateral retained deciduous lateral incisors, with no signs of mobility. [*Figure 1a*] Furthermore, it was observed that all the four mandibular incisors were deciduous with normal physiologic mobility. [*Figure 1b*] A 7mm spacing was found in both arches, between the incisors, with no midline shift. A bilateral Class I molar relationship was noted.

The child was born to non-consanguineous parents. Patient's mother reported a normal pregnancy with vaginal delivery, normal developmental milestones and no history of any trauma/ infections in the past. Family history was non-contributory with the mother reporting no such similar anomaly in the siblings/father/any of the immediate family members. Based on the past history, it was noted that the child was apparently healthy, non-syndromic and not suffering from any systemic conditions.

An orthopantomogram was taken as a part of radiographic investigation (Figure 1c). OPG revealed absence of permanent successors of the maxillary lateral incisors, the mandibular central and right lateral incisors, and absence of the crypt of the mandibular right second molar was also noted whereas crown formation was complete for the opposing and contralateral permanent second molars. The final diagnosis was of oligodontia due to congenital agenesis of 6 permanent teeth. The permanent successors of all the other teeth were present, though the amount of dental development seemed to be delayed considering the chronological age of the child. Objective measure of the dental age was done using the Demirjian's method,[8] considering seven permanent left mandibular teeth from central incisor to second molar. If any tooth was congenitally missing, the contra lateral tooth was considered. Each tooth was assigned a score based on its developmental stage and the sum of seven individual scores gave a maturity score. This score was converted into the dental age using conversion chart. By this method, the child's dental age was estimated to be 7.6 years as opposed to the chronological age of 11 years.

The parents were informed regarding the diagnosis and the prognosis of retained deciduous teeth. Multidisciplinary treatment approach was planned with prosthetic rehabilitation for the missing teeth and orthodontic treatment. The chief complaint was addressed and the decision of keeping the child under observation was made since the parents were not concerned about the developing malocclusion.

Case Report 2

A 10-year-old female reported to our department with the chief complain of missing teeth and malocclusion. There was no history of trauma or decay leading to the loss of teeth. Her medical history was non-contributory. Intraoral clinical examination revealed a 7mm spacing between permanent central incisors due to a peg shaped left lateral incisor and missing right lateral incisor. [*Figure 2a*] The lower central incisors were deciduous with no abnormal mobility. [*Figure 2b*] Further questioning revealed that the father had a history of orthodontic therapy due to multiple missing permanent teeth. The mother's history was non-contributory.

An orthopantomogram was taken as a part of radiographic investigation [*Figure 2c*].OPG revealed absence of the maxillary right lateral incisor and permanent mandibular central incisors. Furthermore, absence of the crypt of

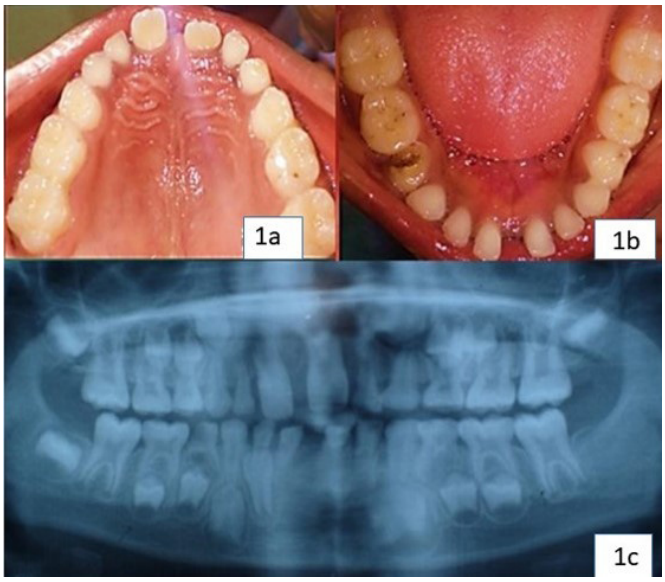

Fig. 1. Case 1 showing (1a) Retained bilateral deciduous lateral incisors in the maxillary arch; (1b) Presence of all four retained lower deciduous incisors; (1c) Orthopantomogram revealing absence of the permanent maxillary lateral incisors, the mandibular central and right lateral incisor, and absence of the crypt of the permanent mandibular right second molar.

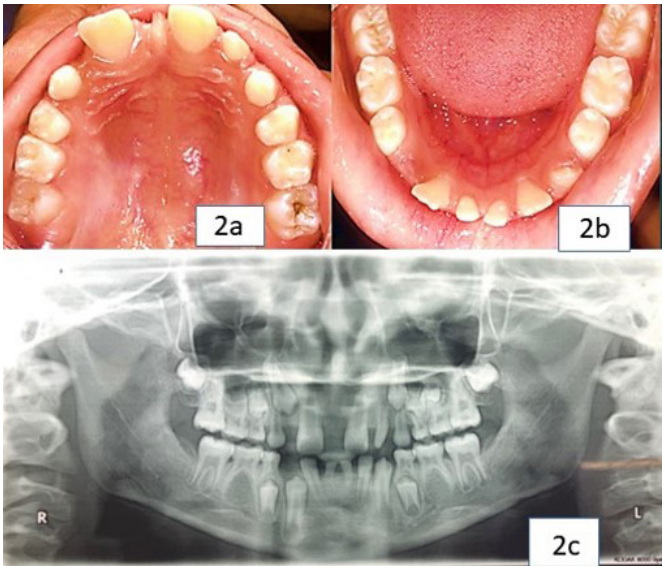

Fig. 2. Case 2 showing (2a and 2b) Retained deciduous mandibular central incisors and 7mm midline diastema in the maxillary arch, peg shaped left lateral incisor and missing right lateral; (2c)Orthopantomogram revealing absence of the maxillary right lateral incisor and permanent mandibular central incisors. Absence of the crypt of the bilateral mandibular second premolar and second molar, and maxillary right premolar also noted.

the bilateral mandibular second premolar and second molar, and maxillary right premolar was also noted. The final diagnosis was of oligodontia due to congenital agenesis of eight permanent teeth. The parents were informed of the condition and the need for a multidisciplinary treatment approach. Following orthodontic consultation, it was decided to treat the child in 3 phases: Phase 1 being orthodontic therapy, Phase 2 being prosthetic rehabilitation of the missing teeth and phase 3 involving maintenance and review.

Discussion

Oligodontia, or the congenital absence of more than six teeth in primary, permanent or both dentitions, is of variable etiology. It may occur as a part of some syndrome or as an isolated, non-syndromic finding. In non-syndromic cases, several etiologic factors like trauma, radiation, infection and metabolic disorders at the time of formation of tooth germ has been suggested [3], which are known to cause mild dysplastic expression of the ectoderm leading to agenesis [9]. Heredity or familial distribution is also seen in cases of congenital agenesis, although the pattern and severity of expression may vary, as confirmed by studies on monozygotic twins [10,11]. Certain genes that play a crucial role in early craniofacial development like those encoding a signaling molecule (TGFA), transcription factors (MSX1 and PAX9), signaling protein AXIN2, and a transmembrane receptor of fibroblast growth factor (FGFR1) are involved in the etiology of human non-syndromic oligodontia [5,12], A unifying etiological theory suggesting a polygenic mode of inheritance, along with the influence of epistatic genes and environmental factors has been proposed [6].

Excluding the third molars, the incidence of missing permanent teeth has been reported to vary from 2.6% to 11.3%. Asian population ranks third in the occurrence of cases of congenital missing teeth, with females often more affected than males [2,13]. Gupta *et al*. [14], in an Indian study, reported that the most frequent anomaly of tooth number was hypodontia, with a prevalence rate of 4.19%. Oligodontia, on the other hand, as in this case, was relatively uncommon with a prevalence of 0.36%.

Arch-wise comparisons have revealed that maxillary are more commonly missing as compared to mandibular teeth [15]. The last teeth of a class are commonly involved (I2, P2, M3-Lateral incisor, second premolar and third molar), suggesting a possible link with evolutionary trends [5,15]. Also, while posterior agenesis might be sporadic, anterior agenesis may depend more on genetic influences [16]. In the first case, a definite etiology for the congenital absence of permanent teeth could not be determined and thus was assumed to be idiopathic. In the second case, however, a familial pattern was noted.

Congenital missing teeth have direct clinical implications. Comprehensive, often expensive treatment is required, depending on the degree of agenesis, ranging from minor prosthodontic interventions like fixed partial dentures to complex orthodontic therapy. It might represent an interdisciplinary challenge, especially since children with oligodontia become aware of their condition at an early age and become more keenly involved in the decision-making process [17,18]. Robinson *et al*. [19] have suggested 4 basic options in the management of retained primary teeth with congenitally missing permanent counterparts: to either retain, retain and modify, extract and manage space either by orthodontic (space closure) or prosthetic means (fixed replacement).

Retained deciduous teeth tend to become ankylosed and in infraocclusion. This could lead to deficient growth of the dentoalveolar component [20]. Mandibular incisor agenesis, in particular, has a pronounced effect on symphysis growth and morphology, with affected patients exhibiting significantly smaller symphysis area than normal patients, resulting in minimal volume of bone for the placement of end-osseous implants at a later period [21]. In both the cases reported had agenesis of the mandibular incisors. Factors like retroclination of alveolar bone, soft tissue imbalances due to loss of lingual support should be taken into consideration during the planning of orthodontic therapy.[3] However, when there is a pre-existing tooth material- arch length discrepancy as in the case of a crowded Class I malocclusion, congenital absence of 1 or more mandibular incisors may be advantageous [22]. Case report 1 and 2 had class I molar relation on both right and left side, therefore in these cases missing teeth would be advantageous if there will be a tooth material arch length discrepancy.

In the case of absence of maxillary incisors, there is a tendency for counter clockwise rotation of the mandible, with shorter upper and lower anterior facial heights resembling those of cleft patients [23]. In such cases, orthodontic therapy should be aimed at protraction of maxillary posteriors, increasing the vertical dimension, and in general prevent worsening of the Class III tendency. In our cases maxillary laterals were missing with angles class I molar relationship on both the sides.

If malocclusion is not a major problem, restorative procedures are indicated to address the aesthetic concerns of the patient. Initially, removable partial acrylic dentures can be given, as rigid fixed prosthesis is contraindicated during active craniofacial growth [17]. Primary teeth can be used as abutments for bridgework if there is satisfactory root length, morphology and coronal structure [19]. Ultimately implant placement may be required. Decreased bone volume may however complicate treatment necessitating prior ridge augmentation or sinus grafting procedures [24].

Conclusion

It is essential that practitioners monitor the developing dentition with establishment of a proper review schedule. Non-eruption of the permanent tooth more than one year later than expected, or even after six months following

the emergence of the contralateral tooth, warrants a high degree of suspicion. A multidisciplinary team, including pediatric, restorative and orthodontic specialists, is advised. In addition, prior to formulation of any treatment plan, due consideration to the general issues such as the patient's systemic and oral health, motivation and expectations should be given.

Author's Contributions

SS (Investigation, Methodology, Writing-original draft, Writing- review & editing)
SRU (Methodology, Writing-original draft, Writing- review & editing)
ANP (Conceptualization, Supervision, Writing- review & editing)
RO (Supervision, Writing- review & editing)
SBS (Supervision, Writing- review & editing)
AR (Supervision, Writing- review & editing)
KYM (Supervision, Writing- review & editing)

Conflict of Interest

The authors of this article declare that there is no conflict of interest.

Informed consent

Patients have signed the written consent for publication of this report.

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