Case Report

NON SYNDROMIC FAMILIAL HYPODONTIA - A CASE SERIES

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ABSTRACT

Aim: The aim of this case report is to discuss the features of non-syndromic familial hypodontia, which was noted in three siblings who had an unremarkable medical history.
Summary: Congenital absence of teeth is a common dental anomaly referring to teeth that fail to erupt in the oral cavity and remain invisible in radiographs. Congenital tooth agenesis is defined by the number of missing teeth, excluding the third molars; hypodontia refers to the absence of less than six teeth, while oligodontia is the absence of six or more teeth.
Tooth agenesis can occur as part of a genetic syndrome such as the cleft lip and palate, ectodermal dysplasia and Trisomy 21 (syndromic hypodontia) or an isolated trait (nonsyndromic hypodontia). Non-syndromic hypodontia is the most common form and can be familial or sporadic. The most supported etiological theory suggests that tooth agenesis may be due to a combination of genetic and environmental factors, which lead to disturbances in the tooth germ during the initial formation stages.
This case series reports bilateral agenesis of the maxillary canines in siblings, which is quite rare in occurrence. The management of these anomalies involves a multidisciplinary approach in order to restore esthetics and function. An early diagnosis can lead to effective treatment planning and satisfactory results.
Key learning points: The clinical manifestations as well as the short and long term management of this condition are discussed.
Keywords: hypodontia, congenital, nonsyndromic, management.

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1. Introduction

Hypodontia is a very common dental anomaly that occurs mainly in the induction and proliferation stage of tooth development, leading to tooth agenesis, posing a significant clinical problem.
It can occur in both the primary and permanent dentition and its classification is based on the number of missing teeth, excluding the third molars [1]. Anodontia is defined as the complete absence of teeth either in one or both arches [1,2]. Oligodontia is a severe and rare form of tooth agenesis which is defined as the absence of six or more teeth, while the congenital absence of less than six teeth is referred to as hypodontia.
Hypodontia can be inherited as an isolated trait or is usually associated with several syndromes such as ectodermal dysplasia, Down Syndrome and cleft lip and palate [2,3].
Non-syndromic hypodontia is more common, with varying numbers of teeth that can be involved; however, the incisor-premolar type predominates [3].
Various studies report that hypodontia is more commonly seen in the maxillary arch and the permanent dentition is much more affected than the primary dentition [3,4].
A review of the literature indicates that the etiology of hypodontia is multifactorial and has been reported to be due to genetic as well as environmental factors [5].
Recent studies indicate the role of the genes in the tooth morphogenesis from the initiation, patterning and histogenesis of dental tissues. Mutations in some of these genes, mainly MSX1, PAX9 and AXIN1 may result in tooth agenesis, although the exact mechanism is not known [6-9]. The environmental factors implicated in tooth agenesis include trauma to the dental region, chemotherapy or radiotherapy and maternal infections during tooth development [6,7].
Prevalence of hypodontia in the permanent dentition, excluding the third molars, is about 4.5 – 7.4% in Caucasians; while including the third molars is about 9-30% [2,4].
A 2016 systematic review and meta-analysis found that there was no difference between the prevalence of hypodontia among the different ethnic groups with each affected Negroid, Mongoloid, and Caucasian having 1.871, 1.900, and 1.889 missing teeth respectively [10].
The most frequently affected teeth are the mandibular second premolar and the maxillary lateral incisor [4]. The congenital absence of canines in the permanent dentition is reportedly a very rare occurrence with prevalence rates varying from 0.06% to 0.18% [11].
The present cases report familial non-syndromic hypodontia in three siblings, of which two cases show bilateral maxillary canine agenesis.

2. Case report

2.1. Case 1

A 7.9-year-old girl (ZA) presented with her father at the department of pediatric dentistry for an initial dental

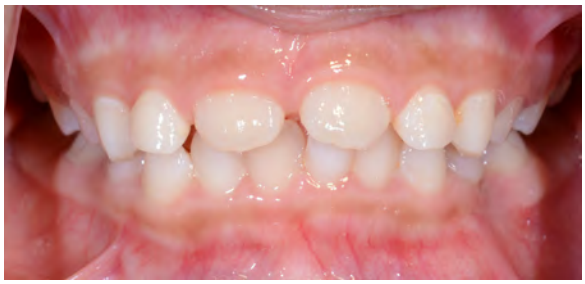

Figure 1. Intraoral frontal view, ZA.

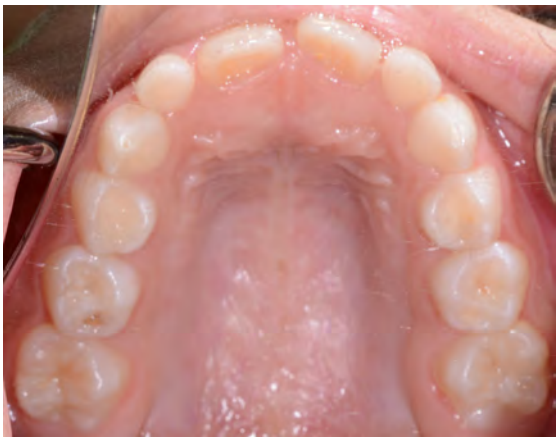

Figure 2. Upper occlusal view, ZA.

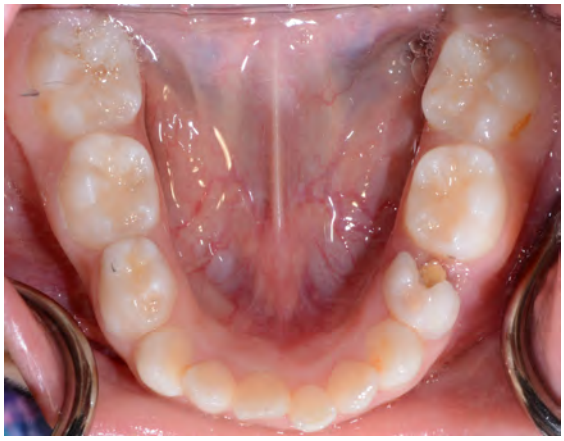

Figure 3. Lower occlusal view, ZA.

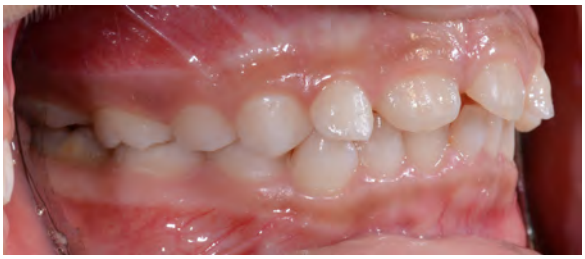

Figure 4. Right buccal view, ZA.

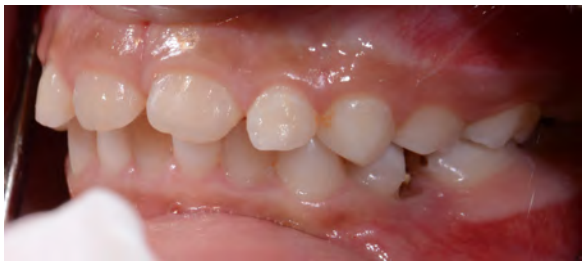

Figure 5. Left buccal view, ZA.

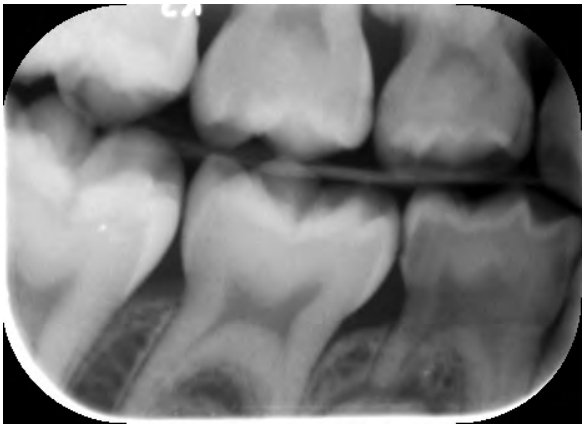

Figure 6. Right bitewing, ZA.

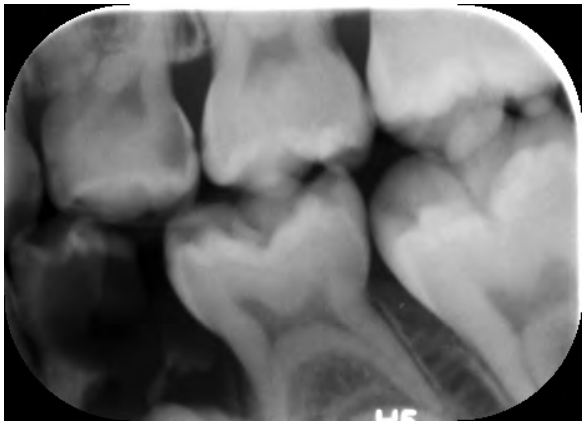

Figure 7. Left bitewing, ZA.

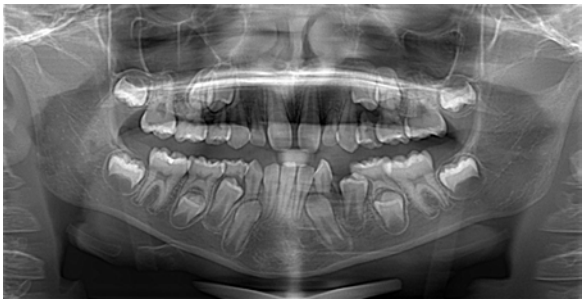

Figure 8. Orthopantomogram, ZA.

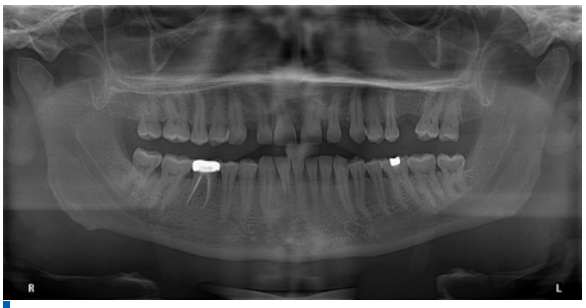

Figure 9. Orthopantomogram, father.

visit with no chief complaints. Her medical history was unremarkable with no systemic conditions or syndromes.

Dental History: An extra-oral examination revealed a convex profile with an increased over-jet (5 mm) and the patient's skin and hair were found to be normal in texture and appearance. The intraoral examination showed the child to be in the mixed dentition stage (Fig. 1-5). Multiple teeth, namely 55, 54, 53, 63, 64, 65, 73, 84 (FDI Notation system) had proximal carious lesions which were detected clinically and confirmed by the initial bitewing radiographs (Fig. 6, 7). Tooth 74 was noted to be non-restorable.

A comprehensive dental treatment plan was formulated and discussed with the parent, which included restoration of the carious teeth with composite restorations and extraction of tooth 74. The patient however did not return back for any treatment for almost one year, despite repeated attempts to contact them.

Upon the child's return for treatment at 9 years of age, a new dental assessment was conducted which included an orthodontic consultation due to the Class II malocclusion with the increased over-jet.

A routine panoramic radiograph was thus taken which revealed agenesis of teeth 13, 12, 22, 23 and 32 (Fig. 8).

The corresponding primary teeth (53, 52, 62, 63) were retained with no radiographic evidence of tooth resorption or mobility. Tooth 72 had exfoliated and teeth 31, 41 and 42 were present.

Based on the history and clinical findings, a diagnosis of non-syndromic hypodontia was made.

A comprehensive treatment plan using a multidisciplinary approach was formulated in order to restore both esthetics and function and discussed with the father.

The short-term plan included restoration of the carious teeth and maintaining the retained primary teeth (53, 52, 62 63) until they showed clinical or radiographic signs of exfoliation on routine follow up appointments. The long term orthodontic plan would then be initiated which would involve extraction of all the retained primary teeth, moving the maxillary premolar teeth (14 and 24) into the respective canine spaces and reshaping them to resemble the maxillary canines (13 and 23).

The space for the missing maxillary lateral teeth would be preserved. Once orthodontic treatment was completed, a resin retained bridge would be fabricated to replace 12 and 22 as a temporary measure until implants could be placed once the child turns 18 years of age.

Due to the diagnosis of the hypodontia, a detailed medical history was obtained from the parent which revealed a non-consanguineous marriage, with no history of genetic conditions in both parents. The child was the third oldest among four children. The

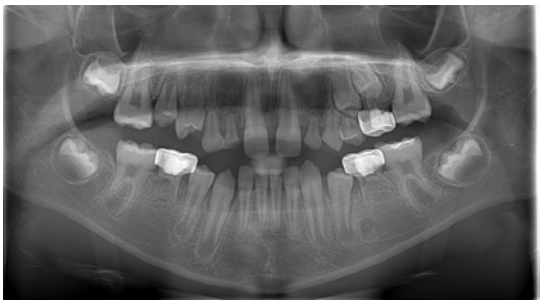

Figure 10. Orthopantomogram, AA.

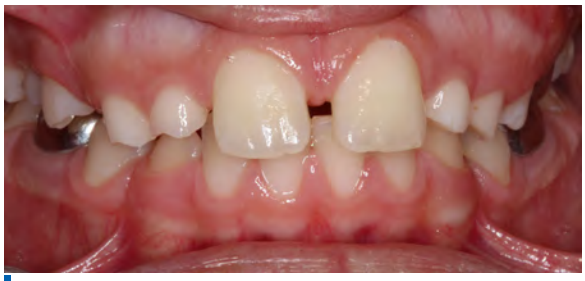

Figure 11. Intraoral frontal view post-op, AA.

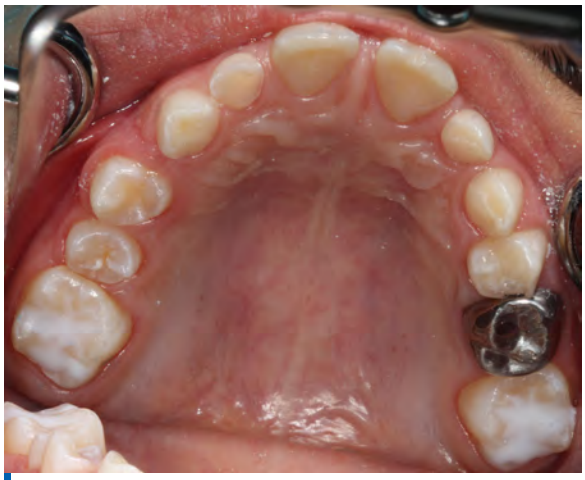

Figure 12. Upper occlusal view post-op, AA.

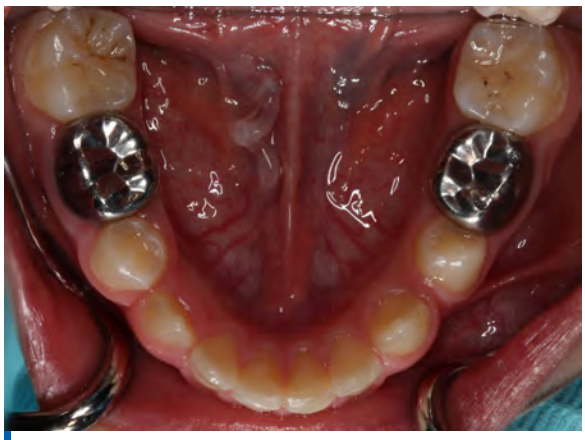

Figure 13. Lower occlusal view post-op, AA.

father agreed to a panoramic radiographic evaluation of himself and was advised to bring in the remaining children for a thorough dental evaluation to detect familial hypodontia.

The father's radiograph showed spacing in the upper arch, no congenitally missing teeth and a history of extraction of tooth 26 (Fig. 9).

The two older children (Cases 2 and 3) however had congenitally missing teeth. The youngest child was 2

years of age and hence no radiographic evaluation was conducted.

2.2. Case 2

AA was 10 years of age with an unremarkable medical history. The panoramic radiograph revealed oligodontia with teeth 13, 12 22, 23, 45, and 55 congenitally missing (Fig. 10).

AA presented with a Class II malocclusion and a 7mm anterior overjet (Fig. 11, 12, 13). A similar short term and long term orthodontic plan was devised to correct the missing teeth, where in teeth 14 and 24 would be positioned in the place of the permanent canines and the space for the lateral incisors, 12 and 22, would be preserved for future implants, once the child turns 18 years of age.

Teeth 75 and 85 would also be extracted and the space would be closed orthodontically.

2.3. Case 3

OA was 12 years of age with an unremarkable medical history and presented with tooth 45 congenitally missing and peg shaped upper lateral incisors (12 and 22) (Fig. 14).

The treatment plan included orthodontics to correct the crowding and retaining the space of tooth 45 for a future implant and reshaping the peg shaped maxillary lateral incisors.

The timing of the diagnosis of hypodontia is important so as to choose an appropriate treatment plan which requires good patient and parent cooperation in order to achieve optimum long-term results.

3. Discussion

Non-syndromic hypodontia is the most common form of congenitally missing teeth and can be familial or sporadic and usually occurs as an isolated trait. It can be inherited in an autosomal dominant, autosomal recessive or an X linked pattern.

The etiology of congenitally missing teeth is reported to be a combination of genetic and environmental factors which can cause disturbances to the tooth germ during the initial formation stages [5,6].

The present cases represent a sporadic, non-syndromic familial form of hypodontia. There was no known history of any genetic syndromes in all three cases.

Congenital missing teeth are defined as those that fail to erupt in the oral cavity and are not visible in radiographs. The diagnosis of tooth agenesis should be made after the 6 years of age excluding the third molar, and after 10 years if evaluating the third molar [13]. A panoramic radiograph is the best means to diagnose the number of missing teeth.

Agenesis of the teeth in our case report was evident in the panoramic X rays and confirmed the diagnosis of hypodontia.

The prevalence of hypodontia ranges from 1.6% to 36.5%, depending on the population studied.

A 2014 systematic review found the overall global prevalence of hypodontia to be 6.4%. Different continents displayed different prevalence values with Africa being the highest at 13.4%, followed by Europe at 7%, Asia and Australia both 6.3%, North America 5% and Latin America and the Caribbean displaying the lowest

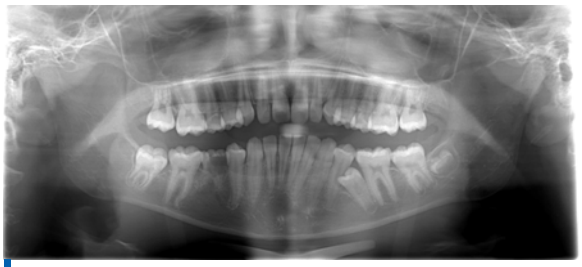

Figure 14. Orthopantomogram, OA.

prevalence values at 4.4%. Most individuals had only one or two teeth missing [3].

Mandibular second premolars are the most commonly affected and account for 29.9% of all congenitally missing teeth, followed by maxillary lateral incisors (24.3%). Maxillary canines on the other hand made up only 2.5% of all congenitally missing teeth [3].

Our case series showed three siblings with variable expressions of hypodontia. Case 1 and 2had congenitally missing maxillary canines in addition to maxillary lateral incisors. The occurrence of bilateral congenitally missing canines is very rare with prevalence rates reported to be as low as 0.14% by Lombardo et al [14].

Hypodontia is often associated with other dental anomalies such as interdental spacing, microdontia, delayed tooth formation, over retained primary teeth, and reduced development of the alveolar bone and taurodontism [15]. Some studies however indicate that the agenesis of permanent teeth shows a strong correlation to the absence of the corresponding primary predecessors, while others indicate that the absence of a permanent successor causes a delay in the root resorption of the corresponding primary teeth [4,12].

Cases 1, 2 and 3 all showed interdental spacing and retained primary teeth where the permanent successors were congenitally missing. Additionally, Case 3 also showed microdontia in the form of maxillary peg shaped laterals.

The treatment approach for hypodontia is specific to each case and is dependent on many factors such as the number of teeth missing, the condition and longevity of the primary predecessor, size and number of teeth remaining in both arches, the occlusal status, facial growth patterns, patient and parent preferences including the financial aspect of the treatment options offered [4,16,17].

A multidisciplinary approach is highly recommended which helps in the short and long-term treatment planning. The various specialists usually involved in the care of individuals with hypodontia include pediatric dentists, orthodontists, prosthodontists and oral surgeons.

In the present cases, initial consultations were done with the orthodontist and prosthodontist. The treatment plan was discussed in detail with the parents and the patient preferences were also taken into consideration.

In cases of hypodontia, the treatment options usually include the timely extraction of the over retained primary teeth that do not have a permanent successor to allow spontaneous orthodontic space closure with or without orthodontic alignment or placement of a prosthetic restoration to replace the missing tooth/teeth. In some cases, the primary tooth is retained to allow growth and impede resorption of the alveolar bone which will help

preserve the volume of the bone. This will facilitate the future placement of an implant in favorable locations, with a reduced need for bone grafting [4,14,16,17].

Cases 1 and 2 displayed little or no resorption of retained maxillary primary canines at the time of diagnosis. The short-term plan included retaining all the primary teeth (53, 52, 62, 63) with the missing permanent successors (13, 12, 22 and 23) as long as possible until they showed signs of tooth resorption. This would allow for alveolar bone growth which would help maintain bone height for future implant placement.

According to GunaShekhar et al [4], the survival rate of primary canines without permanent successors might be long and would provide a functional advantage to patients with hypodontia or oligodontia.

In cases with permanent maxillary canine agenesis, the treatment options include:

Orthodontic movement of a premolar into the canine space and reshaping it to resemble the canine, esthetically and functionally; creating or retaining the adequate space in the area of the canine tooth and placing a prosthetic appliance such as temporary cemented bridges, resin bonded artificial teeth, or removable appliances with acrylic teeth to serve as space maintainers and to improve the patient's appearance. In some of these cases, an implant may be placed in the available space once the child reaches the appropriate age at the end of the facial growth period [18].

In both Case 1 and 2, the maxillary premolar teeth (14 and 24) were orthodontically moved to the missing canine (13 and 23) space and reshaped to resemble maxillary canines. The long-term plan also included preserving the space for the maxillary lateral incisors (12 and 22), placing a temporary resin cemented bridge followed by implant placement in the region once the facial growth period is completed at approximately 18 years of age.

In Case 3, the space for missing tooth 45 was also retained for a future implant and the peg shaped maxillary lateral incisors were built up cosmetically.

The diagnosis of hypodontia should thus be made early which will help making the patients and parents aware of the existing condition as well as help in the treatment planning.

4. Conclusion

Hypodontia is a multifactorial dental anomaly which may occur in isolation or as part of a genetic syndrome. The number of missing teeth varies among individuals and negatively affects esthetics and function. The most commonly missing teeth are the mandibular second premolars and maxillary lateral incisors. The management of this condition is multifactorial and an early diagnosis can lead to effective treatment planning.

Author contributions

Equal contribution to the paper.

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Questions

1. Oligodontia is defined as the agenesis of how many tooth/teeth?

❑a. One permanent tooth;
❑b. One primary tooth;
❑c. Six or more permanent teeth;
❑d. Six or more primary teeth.

2. Which are the most commonly congenitally missing teeth?

❑a. Mandibular first premolar and maxillary central incisor;
❑b. Mandibular second premolar and maxillary lateral incisor;
❑c. Maxillary first premolar and mandibular central incisor;
❑d. Maxillary second premolar and mandibular lateral incisor.

3. Which dental anomaly is usually associated with hypodontia?

❑a. Interdental spacing;
❑b. Over retained primary teeth;
❑c. Taurodontism;
❑d. All of the above.

4. What is the treatment approach for hypodontia dependent upon?

❑a. Number of missing teeth;
❑b. Size of the remaining teeth;
❑c. Occlusal status of the patient;
❑d. All of the above.

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