Bi-Maxillary Hyper-Hypodontia: A Unique Case Report with Review of Literature

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Abstract
The term “hypo-hyperdontia” is used to describe condition in which agenesis of some teeth are seen to occur simultaneously with supernumerary teeth within the same individual. Supernumerary teeth may resemble the permanent teeth sometimes but generally are of different shape and size. These teeth may remain embedded in the alveolar bone may cause disturbance to the developing teeth or can erupt into the oral cavity. Contrary to the presence of extra tooth in arch few complement of the teeth were missing in the arch indicating presence of concomitant hyper hypodontia. The erupted or unerupted tooth might cause aesthetic and/ or functional problems especially if it is situated in the maxillary anterior region. Recently, many genetic studies have been conducted to explore these developmental anomalies. This case reports a unique case of presence of supplemental supernumerary maxillary central incisor with hypodontia of both lower central incisors, which is a rare finding and a literature review, is presented about hypo-hyperdontia in the dentition.

Keywords: Agenesis; Hyperdontia; Hypodontia

Introduction
Development of the tooth is a continuous process with a number of physiologic growth processes and various morphologic stages interplay to achieve the tooth's final form and structure. One such mixed rare numeric anomaly is concomitant hypo-hyperdontia (also known as oligo-pleidontia) characterized by developmental absence of the teeth (hypodontia) and supernumerary teeth (hyperdontia) co-existing in the same individual [1]. It is the only event in which the developing organ must exit the confines of its bony crypt. Interference with the stage of initiation, a momentary event, may result in some or multiple missing teeth or supernumerary teeth. A supernumerary tooth is one that is additional to the normal series and can be found in almost any region of the dental arch [2]. Supernumerary teeth may resemble the teeth of the group it may belong, i.e., molars, premolars, or anterior teeth, or it may bear little resemblance in size and shape to the teeth with which it is associated [3]. Teeth in excess of the normal complement are named according to the location: mesiodens are located on the palatal side of the maxillary central incisors; supernumerary canines and premolars are located at the normal sites for such teeth; Para molars located buccal to first, second and third molars and distomolars (fourth molars) located distal to third molars.

The etiology behind supernumerary teeth is not totally understood. It has been suggested that supernumerary teeth develops from a third molar tooth bud arising from the dental lamina near the permanent tooth bud, or possibly from splitting of the permanent bud itself (dichotomy theory) [4]. Another theory is the hyperactivity theory which suggests that supernumerary is formed as a result of local, independent, conditioned hyperactivity of the dental lamina (dental lamina hyperactivity theory). Other theory may describe the presence of supernumerary teeth is the atavism theory (given by Bateson 1894), i.e.; presence of certain ancestral characters in individuals though have become lost in the evolution [5,6] this theory is not accepted currently. Supernumerary teeth may be classified on the basis of its morphology. The four morphological types of the supernumerary are conical (peg shaped), tuberculate (more than one cusp or tubercle often paired), supplemental (duplication of teeth) and odontoma (hamartomatous malformations further divided to compound and complex composite odontoma). The most common supplemental supernumerary is the maxillary lateral incisor, but supplemental premolars and molars may also occur.

This particular condition is found to have increased incidence in cleft lip and palate patients, cleidocranial dysplasia or Gardner’s syndrome (multiple impacted supernumerary with polyposis of intestine, multiple sebaceous cyst, and osteomas of bone). A tooth can be stated to be congenitally missing when it is clinically and radiographically missing in the dental arch. Absence of less than six teeth is referred to as hypodontia excluding third molars and absence of more than six teeth is referred to as oligodontia excluding third molars [7,8]. Hypodontia may exist in association with peg shaped laterals, delayed formation and eruption of the other teeth, infra-occlusion of the primary molars, impaction of canines or transposition [9,10], may be found in isolation. Based on the region of occurrence in the dental arch hypo-hyperdontia can be anterior or posterior thought case with posterior hyper hypodontia have not been reported yet. Previously case of concomitant hypo-hyperdontia was observed [11–14].

Case Report
A 13 years boy came with his father to Department of Orthodontics and dent facial at Govt. College of Dentistry, Indore (M.P., India) with
the chief complaint of inability to close lips and irregular teeth and wanted its correction. The patient's medical history and family history were noncontributory. In occlusion the patient had a mild class II division 2 incisor relationship in the mixed dentition, with an over jet of 2 mm and an increased overbite of 4.5 mm (Figure 1). The upper dental midline was shifted to the left by 2 mm with the facial midline while the lower dental midline was coincident.

The lower arch was well aligned; however, both lower permanent central incisors were clinically absent and lower left permanent second molar was erupting. The upper arch was severely crowded with a lack of space for both upper permanent canines. The patient was diagnosed of having supernumerary left central incisors due to which crowding was observed in upper anterior segment and lateral incisor have displaced palatally. Deciduous second molars are present in upper and lower left arch with mobility.

On intra-oral periapical radiographic examination supplemental central incisor is seen to have normal morphological appearance with intact lamina Dura and healthy complete root. Orthopantamogram shows erupting second molars except second molar in right lower quadrant, which is missing, and no tooth bud is visible. Lower central incisors and all the third molars are also missing. Total number of missing teeth is seven from the full complement of permanent teeth. This is a unique combination of missing teeth and supplemental teeth (Figures 2 and 3).

**Treatment**

The treatment in the patient was commenced with a full mouth prophylaxis of the patient followed by extraction of the right supplemental central incisor as that was the tooth that leads to elevation of the upper lip leading to the incompetency giving unaesthetic appearance to the patient. The patient was advised to undergo fixed orthodontic treatment for alignment and leveling. Extraction of retained deciduous second molar was performed so that proper eruption pattern could be maintained.

The patient was advised to give follow up after every 6 months to assess the erupting dentition and oral health status. Patient’s guardian was explained about the erupting second molars and problems, which may occur due to their supra-eruption, to prevent it later on a RPD, could be given to maintain it. In future, prosthetic implant can be placed in place of second molar.

The patient was given treatment option of undergoing orthodontic treatment after the extraction of the supplemental tooth, along with which replacement of single lateral incisor can be done (Figure 3a).

**Discussion**

The exact etiology of hyperdontia is not understood. Some hypothesis has been proposed to explain the formation of supernumerary teeth like atavism, dichotomy, and hyperactivity of the dental lamina and the concept of multi-factorial inheritance. Similarly, some of the theories proposed on the tooth agenesis; like disturbances in differentiation, migration and proliferation of neural crest cells was associated with interactions between the epithelial and the mesenchymal cells during the initiation of ontogenesis may be responsible for hypo-hyperdontia. Few cases of bilateral supplemental maxillary central incisors were described in the literature [15-21]. Gibson reported agenesis of third molars along with a supernumerary tooth in maxilla (mesiodens). Missing mandibular permanent centrals with the presence of supernumerary teeth in maxilla were also reported [22,23]. Hypodontia of mandibular permanent centrals with the presence of midline supernumerary tooth in mandible was reported by many authors [24-27].

Supernumerary most commonly found in the maxillary arch is mesiodens followed by mandibular supernumerary premolars. Supplemental incisors are less common and among incisors supplemental maxillary lateral incisors seem to be more commonly found than central incisors when literature is reviewed (Figure 3b). Mandibular second premolars are the most commonly missing teeth after third molars. Hypodontia found in isolation or associated with some other dental anomaly commonly affects the third molars (9-30%) Followed by maxillary lateral incisors, mandibular incisors and mandibular second premolar. Prevalence of hypodontia in permanent dentition may vary from 2.2 -10.1 % excluding the third molars [28-30] while prevalence of 4.2 % was noted by Gupta et al. [31].

Many cases are reported on concomitant hypo-hyperdontia (CHH). A literature review on the hypohyperdontia shows prevalence ranges from 0.002% to 3.1% one epidemiological study on orthodontic population finds incidence of 0.3% in Non-syndrome CHH [32]. Supernumerary are more common in males while hypodontia is more common in females, CHH has been reported to affect both genders equally [33]. The hypodontia and hyperdontia represent opposite ends of the developmental scale of the dentition showing a gender pre-dominance in males (58 %) than in females, with a 1.3:1 ratio.
When two malocclusions were compared, class III subjects showed significantly higher rates. It can be concluded that there is no genetic relation between congenital anomalies and different malocclusions [39].

Hypodontia is inherited as autosomal dominant mode, but occasionally autosomal recessive and X-linked and polygenic/multifactorial models of inheritance have also been reported. There is large number of genes involved in the odontogenic process, so there are higher chances for mutations to disrupt this process. Tooth agenesis is the most common craniofacial malformation with missing one or more number of teeth (hypodontia, oligodontia or anodontia). Hypodontia can be associated with syndromes or may present as an isolated condition. Hypo-hypertonia is rare in isolation and has been associated with over 50 syndromes (orodigito-facial dysostosis, Hallerman Streiff, Cleidocranial dysplasia syndrome, Ellis van Creveld, Downs’s syndrome, cleft lip and palate etc.) [40,41].

Normal development of the tooth germ is appropriately regulated by molecular signaling pathways, if not can give rise to supernumerary/supplemental teeth. These pathways include components of the Hedgehog, FGF, Wnt, TGF and BMP families [42] mutation of genes have been identified by mutational analysis as the major causes of non-syndrome hypodontia (PAX9, MSX1 and AXIN2) [43]. Hypodontia is resulted from haplo-insufficiency of anyone of these and point mutations can cause some or multiple missing teeth. Expression of transcription factors and signaling molecules operating both intracellular and extracellular are guiding tooth development throughout the odontogenic process. BMP4 (transforming growth factor-beta family) and the transcription factors PAX9 (paired box domain) and MSX1 (home box domain), are examples of controlling factors during the odontogenic process. Expression of PAX9 has been found at the sights of tooth development prior to their being any morphological signs of ontogenesis [44].

Mandibular central and lateral incisors show lowest incidence of permanent teeth agenesis, also permanent teeth agenesis of maxillary central incisors, maxillary cuspids and maxillary first molars also rare [45].

Different phenotypic forms in tooth development are the result of different genes involving different interacting molecular pathways, providing an explanation not only for the wide variety of agenesis patterns and also for associations of dental agenesis with other oral anomalies. Different genes involved in human non-syndrome hypodontia includes genes encoding a signaling molecule (TGFA) and transcription factors (MSX1 and PAX9) that are critical during early craniofacial development, but also genes coding for a protein involved in canonical Wnt signaling (AXIN2), and a trans-membrane receptor of fibroblast growth factors (FGFR1) [46]. In a study, frame shift mutation of Pax9 gene (chromosome 14) was identified as responsible for autosomal dominant oligodontia in a large family for four generations. In some of the affected members, maxillary and mandibular second premolars and mandibular central incisors were absent in addition to the lack of permanent molars; although, a normal primary dentition was present [47,48]. Different mutations in the same gene can result in hypodontia or oligodontia, hence these conditions are not fundamentally different. Further, there are evidence to show that more severe the hypodontia, the smaller the Mesiodistal width of the teeth formed, conversely, patients with supernumerary teeth tend to have significantly larger maxillary central and lateral incisors. The size of teeth in case of hypo-hyperdontia is not documented in the literature (Figure 4).

**Conclusion**

It is a unique case of supplemental/supernumerary maxillary central incisor and missing mandibular central incisors along with missing right lower second molar and all third molars. This type of case is not reported in the literature which shows counter teeth are affected. In these cases a wise treatment plan is necessary to alleviate patient’s problems.

**References**


