Case report:

A cocktail of rarities: Case report and a petite review of concomitant hypo-hyperdontia

Prabhawati Praveen Inamdar BDS, MDS, Sangeetha Palanichamy Venkatesh BDS, MDS, MFDRCS (I), Priya Verma Gupta BDS, MDS, PhD

Corresponding Author: sangs_paedo@gmail.com; Mobile: +968-92405713

Dr. Prabhawati Praveen Inamdar

Specialist Pediatric Dental Surgeon in Department of Dentistry, Royal Oman Police Hospital, Qurum, Muscat, Sultanate of Oman.

Funding: The authors declared that this study received no financial support.

Data Availability Statement: Not applicable

Conflicts of Interest: The authors declare that they have no competing interests.

Acknowledgments: Many thanks to Colonel Dr. Nasser Al Manthery, Dr. Sriman Krishnan, and Dr. Khalid @ Royal Oman Police Hospital, Sultanate of Oman, for their valuable opinions and support.

How to cite this Article: Dr. Inamdar. A cocktail of rarities: Case report and a petite review of concomitant hypo-hyperdontia. J Update Pediatr Dent. 2021; 1: (1) 30-38 http://doi.org/10.54276/JUPD.2021.1106

Abstract

A teenager with swelling in the left maxillary canine region exhibited an atypical combination of three rare sporadic anomalies of teeth. The clinical and radiographic evaluation revealed an unerupted odontoma, an impacted maxillary left deciduous supernumerary canine, an impacted maxillary left permanent canine, and the bilateral absence of mandibular lateral incisors. The findings led to the diagnosis of a non-syndromic case of concomitant hypo-hyperdontia. Only a small number of cases of hypo-hyperdontia have agenesis of mandibular lateral incisors and odontomas, but none have deciduous supernumerary canines. The authors thus bring a new combination of hypo-hyperdontia cases. This case report also discusses the treatment options of hypo-hyperdontia and the consequences of delaying treatment.

Keywords: Complex composite odontoma, Supernumerary canine, Hypodontia, Hyperdontia, Hypo-hyperdontia.
Introduction

Agenetic and hypergenetic teeth are numerical developmental odontogenic anomalies. Agenesis of fewer than six teeth is hypodontia, and extra teeth characterize hyperdontia.\(^1\) As with opposites, hypodontia and hyperdontia are two extremes of tooth development that can occur together in the same individual. Currently, such clinical conditions are called concomitant hypo-hyperdontia (CHH). Earlier terms were concomitant hypodontia and hyperdontia,\(^2\) oligopleiodontia,\(^3\) and hypo-hyperdontia.\(^4\) CHH is rare, with a prevalence of 0.002\% to 0.1\%.\(^5\) Taurodontism,\(^7,8\) dens invaginatus,\(^9\) double teeth,\(^10\) and odontoma\(^11\) are anomalies associated with hypo-hyperdontia. More than 50 syndromes are associated with CHH;\(^6\) nevertheless, non-syndromic CHH (NS-CHH) is uncommon.\(^5\)

As individual entities, the prevalence of hypodontia of mandibular lateral incisors,\(^12\) anterior maxillary complex composite odontoma,\(^13\) and permanent supernumerary canines (P-SNC)\(^14\) are relatively rare. The deciduous supernumerary canines (d-SNC) are even rarer finds.\(^15\) The agenesis of mandibular incisors (A-MLI) can cause developmental disturbances of the soft tissues, hard tissues, and occlusion.\(^16-19\) The complex odontomas frequently cause bony expansion,\(^20\) while the SNC usually causes the impaction of the associated permanent teeth.

Thus, the coexistence of the aforementioned single entities in NS-CHH may negatively affect the patients’ esthetics, functionality, and quality of life.\(^21,22\)

Case and age-specific treatment protocols of CHH may require interdisciplinary integration phased over time.\(^23\) Multiple factors influence treatment complexity: number, type, and location of hypo-hyperdontia are a few contributing factors. The anterior complex composite odontoma, A-MLI, SNC, NS-CHH are rare sporadic numeric aberrations of teeth. Their concomitant occurrence in the same individual is unusual; thus, it serves as a prototype. This paper adds to the growing body of work on SNC and NS-CHH.

Case Presentation:

A 13-year-old female and her mother attended the Department of Pediatric and Preventive Dentistry for an upper jaw swelling. The patient's health history, family, and social history were not relevant. At nine years of age, the patient had a history of avulsion of the upper right central incisor. The patient presented with a moderate, bony hard, non-tender intraoral swelling in the left maxillary canine region. There was no extra-oral involvement or facial asymmetry. The maxillary right central incisor left maxillary canine, both the lower permanent lateral incisors were clinically absent (Figure 1).

Dental radiographs revealed an unerupted complex composite odontoma, an impacted supernumerary primary canine, an impacted upper left permanent canine stacked in the maxillary left canine region. Impacted upper left permanent canine was in close approximation to the root apices of the upper left permanent central and lateral incisor with no apparent root resorption. The upper right maxillary central incisor, lower right and left lateral incisors were absent. All other teeth appeared normal for the patient's dental and chronological age [Orthopantomograph Intraoral Periapical Radiograph Figure 2].

Figure 1. Preoperative intra-oral clinical pictures showing the buccal bulge in tooth 23 region.

ISSN: XXXX-XXXX
A cocktail of rarities: Case report and a petite review of concomitant hypo-hyperdontia

Inamdar, 2021

Figure 2. Panromic radiograph and intra-oral radiograph showing presence of odontome in tooth 23 region.

Treatment:

A systematic multidisciplinary management was required. The odontoma and SNC required surgical intervention. An orthodontic and prosthodontic evaluation was necessary to address the impacted canine and the missing teeth. The dental team reviewed clinical and radiographic findings and discussed possible treatment options in concert with the parents.

The parents' financial profile precluded long-term treatment, but the parent-approved immediate surgical removal of the hyperdontic teeth under local anesthesia. A routine preoperative blood test preceded the minor oral surgical procedure. A buccal flap was raised, surgical extraction of OD and d-SNC was performed [Figure 3]. 3-0 silk sutures placed and excised odontomas were shown in [Figure 4].

Figure 3. Surgical removal of odontoma in tooth 23 region.

Figure 4: Excised odontomes
Follow-Up:
Preoperative healing was uneventful following suture removal

Table 1: Documented cases of non-syndromic CHH with missing mandibular lateral incisor

<table>
<thead>
<tr>
<th>Authors</th>
<th>year</th>
<th>Age/ Gender</th>
<th>Hyperdontia</th>
<th>Agenesis</th>
<th>other significant findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Matsumoto et al [61]</td>
<td>2001</td>
<td>8/F</td>
<td>22,25</td>
<td>32</td>
<td>-</td>
</tr>
<tr>
<td>Anthonappa et al [5]</td>
<td>2008</td>
<td>5/F</td>
<td>Mesiodens</td>
<td>72,32</td>
<td>-</td>
</tr>
<tr>
<td>Nayak et al [38]</td>
<td>2010</td>
<td>28/M</td>
<td>Mandibular mesiodens</td>
<td>32, 42</td>
<td>-</td>
</tr>
<tr>
<td>Sharma A [39]</td>
<td>2012</td>
<td>8/M</td>
<td>Mesiodens</td>
<td>32,42</td>
<td>Fusion of 72,73 and 82,83</td>
</tr>
<tr>
<td>Gupta and Popat [57]</td>
<td>2013</td>
<td>11/M</td>
<td>Mesiodens</td>
<td>32,42</td>
<td>-</td>
</tr>
<tr>
<td>Wang et al [38]</td>
<td>2018</td>
<td>7Y3M / M</td>
<td>Mesiodens</td>
<td>32,42</td>
<td>Molar taurodontism</td>
</tr>
<tr>
<td></td>
<td></td>
<td>5Y11M / M</td>
<td>Mesiodens</td>
<td>82,42</td>
<td>72,73 fusion</td>
</tr>
<tr>
<td></td>
<td></td>
<td>7Y8M / M</td>
<td>Mesiodens</td>
<td>32</td>
<td>-</td>
</tr>
<tr>
<td></td>
<td></td>
<td>5Y11M / M</td>
<td>1 (Mesiodens)</td>
<td>82,42</td>
<td>72,73 fusion</td>
</tr>
<tr>
<td></td>
<td></td>
<td>7Y4M / M</td>
<td>1 (Mesiodens)</td>
<td>32</td>
<td>72,73 fusion</td>
</tr>
<tr>
<td></td>
<td></td>
<td>8Y4M / M</td>
<td>2 (Mesiodens)</td>
<td>42</td>
<td>-</td>
</tr>
<tr>
<td></td>
<td></td>
<td>5Y9M / M</td>
<td>2 (Mesiodens, 21 region)</td>
<td>32,42</td>
<td>72,73 fusion; 82,83 fusion; premolar central cusps</td>
</tr>
<tr>
<td></td>
<td></td>
<td>9Y3M / F</td>
<td>1 (Mesiodens)</td>
<td>32</td>
<td>Odontome in 23 region</td>
</tr>
</tbody>
</table>

Wang et al [38] 2018

Table 2: Nonsyndrom concomitant occurrence of with hyperdontia in the canine region

<table>
<thead>
<tr>
<th>Authors</th>
<th>Year</th>
<th>Age/ Gender</th>
<th>Hyperdontia</th>
<th>Agenesis</th>
<th>Any other significant findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Manjunatha et al [76]</td>
<td>2011</td>
<td>26/M</td>
<td>Dilated odontoma w.r.t 23</td>
<td>31,41</td>
<td>Retained 71,81</td>
</tr>
<tr>
<td>Sebastian et al [8]</td>
<td>2013</td>
<td>30/F</td>
<td>Dilated odontoma of peg shaped 12,22</td>
<td>13, 23 and 47</td>
<td>Peg lateral-12,22</td>
</tr>
<tr>
<td>Cholakova et al [10]</td>
<td>2016</td>
<td>15/F</td>
<td>Multiple Odontoma w.r.t 23</td>
<td>35,45</td>
<td>Impacted Canine</td>
</tr>
<tr>
<td>Current case</td>
<td>2021</td>
<td>13/F</td>
<td>Complex composite odontoma in relation to 23 Primary left maxillary supernumerary canine</td>
<td>32,42</td>
<td>Impacted canine above the maxillary left central and lateral incisor</td>
</tr>
</tbody>
</table>

Discussion

Developmental abnormalities of teeth can result from disturbances in the developing dental lamina of the tooth. These include hyperdontia, hypodontia, or morphologic aberrations. The mode of inheritance determines whether the anomaly is syndromic or isolated. Recessive inheritance, new mutations, or stochastic events contribute to sporadic occurrences. Hypodontia is one of the most common developmental problems in children. It can be sporadic, a component of a syndrome, or familial and non-syndromic. Over 80 syndromes are associated with hypodontia. Sporadic anodontia and oligodontia are rare, while sporadic hypodontia is relatively common. The prevalence of hypodontia varies from 0.03% to 10.1% among various populations, with a prevalence of 1% to 4% in Indians. The permanent mandibular incisors show the lowest rate of agenesis. However, their bilateral absence, particularly the mandibular lateral incisors, is highly prevalent in the mongoloid with a female preponderance. Mandibular tooth agenesis may be associated with retained/infraoccluded primary, impacted teeth, and nerve tissue/supporting tissues/oral mucosa disturbances. Mandibular incisor agenesis has a large effect on the mandibular symphysis growth and morphology. Their bilateral agenesis could result in unequal lip-tongue pressure, wide spacing in the anterior region with or without the absence of...
dentition, as well as Class II Div I malocclusion with a severe anterior deep bite. All of these can impact the patients’ functionality and esthetics\textsuperscript{[13,16-19]} The coexistence of A-MLI and hyperdontia is uncommon. Therefore, only a couple of authors have described NS-CHH in conjunction with A-MLI; contrariwise, Wang et al. \textsuperscript{[38]} have documented most instances (Table 1).

Hypergenesis of teeth can be sporadic or associated with systemic syndromes. It can be an essential clue for early diagnosis in some specific disorders.\textsuperscript{[24]} The Odontomes (ODs), and the supernumeraries (SPN)s are classified as two discrete entities, though they may be expressions of the same pathological process.\textsuperscript{[39]} The ODs are benign, odontogenic tumors that develop from the tooth germ or the teeth because of trauma, infection, or a genetic mutation during the growth phase. ODs usually stop growing when the tissues that make up the tumor have fully mineralized. ODs are two main types: compound and complex, although mixed forms have been described.\textsuperscript{[39]} The dilated odontomes (invaginated or gestant) are extreme variants of dens invaginatus.\textsuperscript{[8,12]} Complex ODs predominate in females, whereas compound ODs are more common in males. In the anterior segment of the jaws, most of the ODs are of the compound composite type (61\%), whereas those of the posterior segment are complex composite (34\%). Notably, both types of ODs were more common on the right side of the jaw (compound 62\% and complex 68\%).

In contrast, this case reports a complex composite OD located in the left anterior maxillary region. The radiographic examination is an effective clinical aid in the radiographic diagnosis of the types of ODs. However, histopathology is confirmatory. Retained deciduous teeth, malpositioning or malformation of adjacent teeth, aplasia, devitalization of adjacent teeth, unerupted or impacted teeth, and swelling are the sequel of ODs. Unlike compound ODs, complex ODs cause slight or marked bone expansion with or without evidence of infection.\textsuperscript{[39-46]} ODs and agenetic teeth seldom coexist; thus, only three authors have described NS-CHH with ODs. (Table 2).

Hypergenetic teeth of the canine region are called supernumerary canines (SNCs). P-SNCs are normally rare (0.04 - 4.17\%).\textsuperscript{[14,47]} finding d-SNCs are even rarer.\textsuperscript{[15,48,49]} A majority of SNCs occur in the maxilla unilaterally with supplemental morphology.\textsuperscript{[14,47,50]} The SNCs are associated with a high percentage of mechanical- obstructive pathology such as impaction, delayed eruption, ectopic eruption, root resorption of adjacent teeth, fusion with adjacent teeth, crowding, and cystic lesions. Over half of SNCs (53.85\%) caused the impaction of the permanent canine analogous to the current case.\textsuperscript{[14,15,21,22,47-50]} Stafne\textsuperscript{[51]} stated that denticles and odontomas were found more frequently in the canine region than SNC. Nevertheless, OD and SNC have co-occurred in the current case of NS-CHH. In addition, the SNC has characteristics of a deciduous canine (color, crown morphology, and root length). Prior publications of NS-CHH have not reported deciduous or permanent SNCs. CHH is an unusual mixed numeric anomaly in which the absence of teeth occurs in tandem with extra teeth.\textsuperscript{[5,6,52]} Environmental factors, genetics, or both may cause CHH.\textsuperscript{[5,53]} It is more likely in men than in women, with a ratio of 1:3.\textsuperscript{[5,11,54-56]}

Traditionally, CHH was classified into three major types:\textsuperscript{[54]}
1. The maxillary (maxillary arch alone),
2. The mandibular (mandibular arch alone), and
3. The bimaxillary (maxillary and mandibular arch).

The CHH can further be categorized based on its arch location:

a. Anterior (hypo-hyperdontia involving only anterior region),
b. Posterior (hypo-hyperdontia involving only posterior region), and
c. Anteroposterior (hypo-hyperdontia involving both anterior and posterior region).

Anterior involvement is the most common, followed by the anteroposterior involvement. The posterior hypo-hyperdontia is rare and has never been described before.\textsuperscript{[5,54-56]} Hypodontia in the posterior region and hyperdontia in the anterior region are CHH characteristics. Agenesis of mandibular second premolars occurs more often than maxillary lateral incisors in CHH. Likewise, maxillary mesiodens are more common than mandibular in CHH\textsuperscript{[5,11,7,54,56,58,59]} The literature describes various clinical manifestations of CHH, and this paper adds a notable case to this list. Compared to other documented cases of CHH, this case has an atypical combination of CHH.

Agenetic and hypergenetic teeth in CHH can cause aesthetic and functional issues. Systematic planning and early intervention are necessary to address them. The patient's age and clinical complexity chiefly determine the treatment strategies of CHH. Simple cases may only need minimal intervention, while complex cases may require phased interdisciplinary integration. Treatment planning depends on many factors, including the presenting complaint, patient opinion, patient profile, missing teeth, number of retained teeth, interocclusal space, bone health, size/shape of remaining teeth, jaw size, and edentulous space location\textsuperscript{[9,61-64]}
Multidisciplinary management of complex cases of CHH in phases:

Phase 1: Removal of hypergenetic teeth

The optimal time for removal of the unerupted hypergenetic teeth is debated.[5] In 45% of patients with the extraction of the OD/SPN, the unerupted tooth emerges.[65] Hence, immediate removal is encouraged when SPNs and ODs cause inhibition or delay of eruption, the displacement of the adjacent tooth, interference with orthodontic appliances, or the presence of a pathologic condition. Surgical removal of the SPNs in the deciduous dentition is discouraged as it risks displacing the permanent tooth during the procedure. Some authors suspend the surgical intervention until the root development of the associated permanent successor is complete. In contrast, others await one-half to two-thirds of the root completion to minimize their probability of surgical injury.[9,61,66-68]

Phase 2: Treatment of impacted teeth and hypodontia

Treatment of impacted teeth: The SPNs and ODs most often cause the impaction of associated teeth. Several factors influence the prognosis of an impacted tooth, such as patients’ age, tooth morphology, jaw location, and the amount of arch space.[60] The emergence of impacted teeth may not occur spontaneously after removing SPNs or ODs;[60] in such circumstances, orthodontic treatment of impacted teeth should follow the removal of hypergenetic teeth.[12,47,65,66]

Treatment approaches for hypodontia:

a. No treatment - If arch alignment and esthetics are favorable.[57]

b. Orthodontic space closure
   1. With eruption guidance in mixed dentition.
   2. Before esthetic restorative treatments.

c. Orthodontic space opening
   1. Followed by prosthetic treatment (removal/fixed).
   2. Followed by auto-transplantation.
   3. For implant-supported restorations.[71]

In this case, the odontoma in the maxillary left canine region and the impacted maxillary left d-SNC inhibited and deflected the eruption of the maxillary left permanent canine. The maxillary left permanent canine had not reabsorbed the roots of maxillary left central and lateral incisors. Phase one treatment comprised immediate surgical removal of the odontoma and d-SNC. An orthodontic and prosthodontic consultation was advocated to address the missing maxillary right central incisor, impacted canine, and missing lower lateral incisors. The parent denied further treatment even after being briefed about the impacts of treatment suspension.

Consequences of the lack of or delay in the treatment of CHH:

- Displacement of adjacent teeth,
- Internal and/or external resorption of impacted and adjacent teeth,
- Loss of vitality of impacted and adjacent teeth,
- Ankylosis, transmigration, ectopic eruption,
- Odontogenic cyst and tumors,
- Recurrent pain, referred pain,
- Periodontitis and infections,
- Dento-facial compensation,
- Esthetic, masticatory and phonetic compromise

Observations for dental professionals:

a) Mandibular incisor agenesis causes developmental disturbance of soft tissue, hard tissue, and occlusion.

b) Differential diagnosis (DD) of hypergenetic teeth: ODs, SPNs, dens invaginatus, gemination, fusion, etc.

  c) An extreme variant of dens invaginatus is termed dilated odontome or invaginated odontoma or gestant odontoma.

d) DD of ODs: Radiographic examination is a clinical aid; whereas histopathology is confirmatory.

e) Unerupted SPNs and ODs usually cause the impaction of the associated permanent teeth.

f) The P-SNCs are rare; d-SNCs are extremely rare

Conclusion:

- CHH calls for prompt diagnosis and early intervention.
- CHH largely mandates a well-timed, tailored, multidisciplinary management.
- Mitigate potential damages of CHH.
- Abate deferrals of referrals.

The Authors

Prof. Prabhawati Praveen Inamdar, Professor and Head in the Department of Pedodontics and Preventive Dentistry, Al Ameen Dental College, Bijapur, Karnataka, India
Dr. Priya Verma Gupta,
Affiliation: BDS, MDS, PhD, FPFDA, Professor and Head in
Department of Pedodontics and Preventive Dentistry, Bhabha
Institute of Dental Sciences, Bhopal, Madhya Pradesh, India

References

1. Al-Ani AH, Antoun JS, Thomson WM, Merriman TR, 
Farella M. Hypodontia: An Update on Its Etiology, 
2017;2017:9378325.
2. Camilleri GE. Concomitant hypodontia and 
3;123(7):338-9.
1970;129:309.
5. Anthonappa RP, Lee CK, Yiu CK, King NM. 
Hypohyperdontia: literature review and report of seven 
6. Varella M, Arrieta P, Ventureira C. Non-syndromic 
concomitant hypodontia and supernumerary teeth in an 
Dec;31(6):632-637.
7. Nirmala SV, Sandeep C, Nuvvula S, Mallineni SK. 
Mandibular hypo-hyperdontia: A report of three cases. 
triad: Bilateral dilated odontoma, hypodontia and ped 
9. Segura JJ, Jiménez-Rubio A. Concomitant 
hypohyperdontia: simultaneous occurrence of a 
mesiodens and agenesia of a maxillary lateral incisor. 
10. Cholakova R, Chenevich I, Jordanova S, Oncheva D, 
Chenevich L. A case of compound maxillary odontoma 
and mandibular hypodontia. J of IMAB. 2016 Jul-Sep; 
22(3):1217-1220.
11. Mallineni SK, Jayaraman J, Yiu CK, King NM. 
Concomitant occurrence of hypohyperdontia in a 
patient with Marfan syndrome: a review of the literature 
Nov;3(4):253-257.
12. Phillipsen HP, Reichart PA, Praetorius F. Mixed 
odontogenic tumours and odontomas. Considerations 
on interrelationship. Review of the literature and 
presentation of 134 new cases of odontomas. Oral 
13. Bozga A, Stanciu RP, Măncu D. A study of prevalence 
2014(7):551-554.
14. Türkahraman H, Yılmaz HH, Cetin E. A non-
syndrome case with bilateral supernumerary canines: 
report of a rare case. Dentomaxillofac Radiol. 2005 
Sep;34(5):319-2.
15. Mukhopadhyay S, Mitra S. Anomalies in primary 
dentition: Their distribution and correlation with 
Jan;5(1):139-43.
16. Buschang PH, Julien K, Sachdeva R and Demirjian A. 
Childhood and pubertal growth changes of the human 
17. Fukawa A. Two class II, division I patients with 
congenitally missing lower central incisors. Am J 
18. Endo T, Yoshino S, Oozu R, Kojima K, Shimooka S. 
Association of advanced hypodontia and craniofacial 
morphology in Japanese orthodontic patients. 
19. Endo T, Oozu R, Kojima K, Shimooka S. Congenitally 
missing mandibular incisors and mandibular symphysis 
20. Satish V, Prabhadevi MC, Sharma R. Odontome: A 
21. Hattab FN, Yassin OM, Rawashdeh MA. 
Supernumerary teeth: Report of three cases and review 
of the literature. ASDC J Dent Child. 1994;61(5-6):382-
393.
22. Khandelwal P, Rai AB, Bulgunawar B, Hajira N, 
Masih A, Jyani A. Prevalence, Characteristics, and 
Morphology of Supernumerary Teeth among Patients 
Visiting a Dental Institution in Rajasthan. Contemp 
23. Kariya PB, Singh S, Kulkarni N, Arora A. Bimaxillary 
concomitant hypohypeordontia in a 10-year-old child. 
24. Klein OD, Oberoi S, Huysseune A, Hovorakova M, 
Peterka M, Peterkova R. Developmental disorders of 
25. Guttal KS, Naikmasur VG, Bhargava P, Bathi RJ. 
Frequency of developmental dental anomalies in the 
26. Mostowska A, Biedziak B, Jagodziński P. Novel 
MSX1 mutation in a family with autosomal-dominant 
hypodontia of second premolars and third molars. Arch 
27. Mattheeuws N, Dermaut L, Martens G. Has hypodontia 
increased in caucasians during the 20th century? A 
28. Cunha K, Naikmasur VG, Bhargava P, Bathi RJ. 
Frequency of developmental dental anomalies in the 
29. Vahid M, Vahid B, Vahid M, Vahid M. Prevalence of 
dental anomalies in pretreatment orthodontic patients in 
Western Maharashtra, India: An epidemiological study. 
H, Kothariya R. Prevalance of dental anomalies among 
31. Mallineni SK, Momiuddin A, Patil AK, Kodali RP. A 
Unique Presentation of Concomitant Hypo-
Hypodontia in Seven Year Old Child: A Rare Report. 


