

## Non-syndromic familial hypodontia: rare case reports and literature review

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### Abstract

Congenital absence of the teeth, affecting both physiognomy and mastication, can have a great impact on patients' quality of life. It may appear unilateral or bilateral; frequently, it associates with certain general conditions. Familial hypodontia in clinically healthy patients is rare. Genetic transmission, as a determinant factor in missing teeth, can be autosomal dominant, recessive, or related to the sex chromosomes. In case of congenitally missing permanent teeth, the corresponding deciduous teeth can be still found on the arch at adult age. The aim of this article is to present rare cases of familial non-syndromic unilateral and bilateral hypodontia in Romanian adult siblings, highlighting also a cross gene transmission between aunt and niece and evaluating the treatment options in accordance with patient's age and oral status. It shows that early diagnosis of hypodontia is crucial for the patient's future oral health. The decision to keep the temporary teeth or to extract them is influenced by the presence and status of the deciduous teeth, patient's access to treatment and parent's agreement. Asymptomatic adult patients, in which hypodontia was diagnosed during a routine control, do not usually solicit therapeutic intervention until the deciduous teeth are also lost; still, in situations where complications arise through their loss, the treatment of hypodontia is complex, involving a close collaboration between a team of specialists. The article also includes a detailed review of literature referring to the prevalence of hypodontia among different populations.

**Keywords:** familial non-syndromic unilateral and bilateral hypodontia.

### Introduction

Changes in habitat and cultural environment have led to significant changes in the human diet and the consistency of the consumed food, which determined a decrease in the dental arches size and also in the number and dimensions of the teeth. The absence of one or more teeth can appear as a pattern within families, involving one to six teeth (hypodontia), six or more teeth (oligodontia) or all of them (anodontia). Anodontia as a single condition is usually rare [1] and frequently associated with general pathology. There are a number of general conditions in which anodontia is associated: Pierre Robin sequence, hypohidrotic ectodermal dysplasia, hair-nail-skin-teeth dysplasia, Down's syndrome, palate ectodermal dysplasia syndrome [2–4]. Hypodontia, like anodontia, is often associated with general disorders, but there are rare cases of non-syndromic hypodontia in clinically healthy patients. Factors that cause hypodontia are not fully understood, but some authors showed that there are environmental factors that may favor this disorder [5, 6]. Also, infection, certain drugs, local radiotherapy and trauma can determine changes in teeth development. Genetic transmission may be a determining factor in the occurrence of missing teeth [7]. Congenitally missing teeth

within families can belong to the primary or permanent dentition. Usually, genetic transmission is autosomal dominant, but there are reported cases of autosomal recessive or related to the sex chromosome [8, 9]. If hypodontia is transmitted autosomal dominant, about 50% of siblings will be affected and if it is transmitted autosomal recessive, about 25% of siblings will be affected [10]. Studies in the literature have shown that certain types of genes, such as paired box 9 (*PAX9*) [11], axis inhibition protein 2 (*AXIN2*) [12] and msh homeobox 1 (*MSX1*) [13] may be involved in the congenitally missing teeth etiology. It has been shown that each gene predisposes to a certain category of missing teeth; the *MSX1* gene is involved in the case of missing premolars, and the *PAX9* gene is involved in the case of missing molars [14]. Another gene whose point mutation generates congenitally missing second premolars and third molars is msh homeobox 9 (*MSX9*) [15]. Other genes involved in dental development, such as distal-less homeobox (*DLX*), LIM homeobox (*LHX*) and, recently, interferon regulatory factor 6 (*IRF6*), transforming growth factor alpha (*TGFA*), fibroblast growth factor receptor 1 (*FGFR1*) [16] were studied, the mutation of which can cause changes in the number of teeth. Regarding the non-syndromic hypodontia, it was shown that the genes involved in its transduction are

*AXIN2*, ectodysplasin A (*EDA*), latent transforming growth factor beta binding protein 3 (*LTBP3*) and Wnt family member 10A (*WNT10A*) [16, 17]. Genetic transmission of missing teeth can cause either the absence of the tooth involved, or its presence, but in a small size, typically in a conical form. These teeth have frequent changes in the form and shape of the roots as well, with a reduced root size [18, 19]. The type and location of the congenitally missing teeth varies with ethnicity, excepting the third molar; its absence varies from only one to all four. At the same time, a large morphological variety of the third molars has been observed, small sized ones having a more frequent occurrence. Following the third molar, the second most frequent congenitally missing tooth is the second premolar [20]. In European peoples, mandibular and maxillary premolars are most frequently involved in hypodontia [21]. Studies have shown that the Japanese were generally unaware of the congenitally absence of their mandibular central incisors [22], while in the American people the maxillary lateral incisors [23] were more frequently missing. In this article, three clinical cases of patients belonging to the same family, two adults and one child, with missing teeth and no other associated general illnesses are analyzed in order to highlight a cross-genetic transmission of hypodontia among family members.

## Case presentations

### Case No. 1

A 40-year-old male patient came to our Office in Emergency, accusing pain in the superior right third molar (1.8) area. No general health issue was reported. Intraoral



**Figure 2 – (a and b) Midline shift, minor crowding, missing central mandibular incisors, bilateral canine crossbite and an ectopic first premolar disturbing the occlusion in the right part of the mouth.**



**Figure 3 – Panoramic radiograph: maxillary lateral deciduous incisor with talon cusp, left mandibular third molar with reduced size and modified form.**

examination revealed multiple caries, one located on the upper right third molar, but also a rare situation of missing bilateral permanent mandibular central incisors (3.1 and 4.1) and maxillary lateral incisors (1.2 and 2.2) (Figure 1, a and b). Also, we observed the presence of one deciduous upper lateral incisor on the left side (6.2), and of a temporary mandibular central incisor located on the median line, between the two permanent lower lateral incisors (Figure 2, a and b). The deciduous left maxillary lateral incisor presents a talon cusp, also visible on the panoramic radiograph. The maxillary third molars are present; on the inferior jaw, the left one (3.8) has a different shape and smaller size than normal, while the right one (4.8) is absent (Figure 3). Spaces created by the hypodontia produced midline shift, multiple rotations, and a disharmonious and unstable occlusion through bilateral crossbite, multiple premature contacts and interferences; for example, the permanent maxillary right premolar is erupted in an ectopic position, rotated by more than 90° and in crossbite. No attempt of orthodontic treatment was reported in the antecedents; at presentation, the patient declared that he is interested only in emergency and restorative procedures; he mentioned that his daughter and sister also presented missing teeth and solicited a routine check-up for his 12-year-old daughter.

### Case No. 2

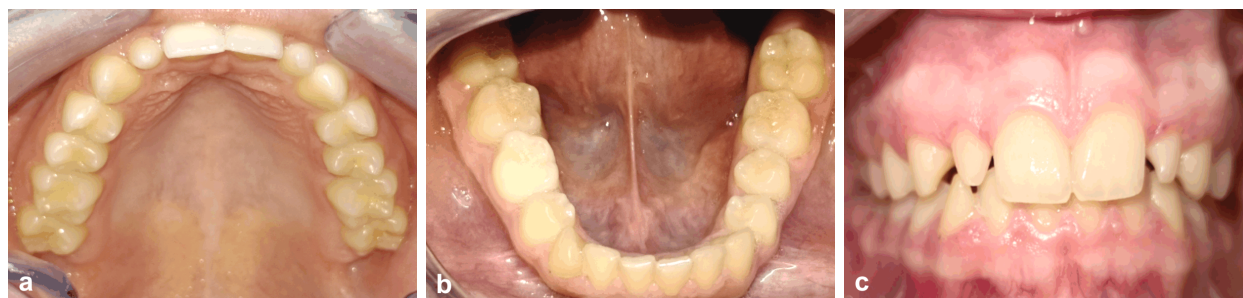
A 12-year-old girl, the daughter of the first patient, came to the Office for a clinical examination, presenting an anterior panoramic radiograph made three years earlier. Clinical and radiological examinations were used to detect any congenitally missing teeth, after the confirmed father's hypodontia.

**Figure 1 – (a and b) The maxillary arch: absence of permanent maxillary lateral and mandibular central incisors, presence of deciduous left lateral incisor with talon cusp on the maxillary arch, presence of one deciduous central incisor on the mandible, multiple rotations.**

The absence of the second inferior premolar on the right side (4.5), with the presence of the deciduous molar still on the arch (8.5), and the modified (conical) shape of the maxillary permanent lateral incisors were noticed (Figure 4, a–c). The mesio-distal dimension of the second deciduous molar, larger than the second premolar, determined the moderate rotation of 4.3; still, there was no midline shift. The mandibular central incisors were on the arch, with no shape-related changes. Also, none of the four third molars were present (Figure 5). Clinical and radiological examination of her mother revealed that she had absolutely no change in the dental formula, or in the size or shape of the teeth. No associated pathology was

present in the girl's case; still, the patient's parents reported that, at the age of three and a half, the girl underwent a congenitally inguinal hernia surgery, the same as her paternal aunt (Figure 6). In her case, the treatment option

agreed with the parents was to keep the deciduous second molar, since it was not affected by caries, for as long as possible on the arch and to reevaluate the possibility of an implant-prosthetic restoration later, in adult age.



**Figure 4 – (a–c) Conical shape of the maxillary permanent lateral incisors; the presence of the deciduous molar on the arch (8.5); minor rotation of permanent right inferior canine (4.3).**



**Figure 5 – Older panoramic radiograph of the patient aged 9: none of the third molars are present.**

Stimată Colegă / Stimat Coleg,

Pacientul(a) [redacted] născut(ă) pe data [redacted] cu domiciliul în localitatea [redacted] nr. [redacted] bloc [redacted] ap. [redacted] sector [redacted] CNP [redacted] a fost internat(ă) în secția CHIR. PEDI. perioada 28.06 - 30.06.2010 cu diagnosticul

**HERNIE INGHINALĂ DREAPTĂ**

Probe biologice: H<sub>2</sub> = 13,3 g/dl, Hct = 38,9%, L = 4,470 /mm<sup>3</sup>  
 TS = 213<sup>u</sup>. Semnele unui - în limite normale

Explorări imagistice:

Medicație administrată: [redacted]

Anestezie: AG - anestezia lorazepam Doctor: [redacted]

Echipa operatorie: [redacted]

Intervenții chirurgicale: CURĂ RĂSINĂ A HERNIEI (29.06.2010)

**Figure 6 – Medical letter for the general practitioner, after the surgery for congenitally inguinal hernia.**



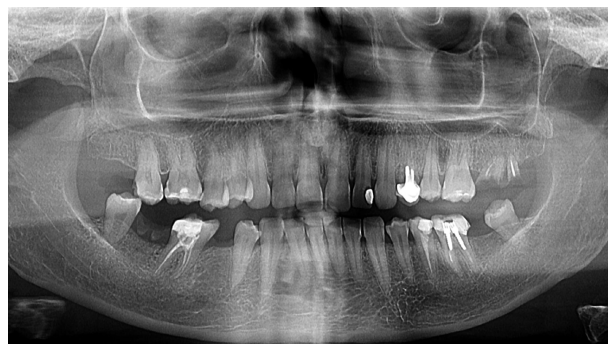
**Figure 7 – (a–c) Permanent maxillary lateral incisors of normal shape and size; absence of the permanent second inferior premolar on the right (4.5), deep bite with minor sagittal inoclusion.**

### Case No. 3

The 44-year-old first patient's sister came to our Clinic for a routine examination and solicited a complete oral rehabilitation. The clinical and radiological investigation showed the permanent maxillary lateral incisors of normal shape and size, the absence of the permanent second inferior premolar on the right (4.5) and deep bite with minor sagittal inoclusion; the patient declared that she lost her deciduous molar (8.5) by decay at the age of 19 years old and refused the restoration of the dental arch at that moment, considering that the 3 unit bridge proposed by the dentist at that time was a too invasive dental procedure (Figure 7, a–c). Similar to her niece, this tooth was the only one involved in hypodontia (Figure 8). Compared to her brother, both upper lateral incisors and lower central incisors are present and have no shape changes. The two lower third molars are present and have normal size and shape; on the maxillary arch, the left third molar (2.8) is damaged by decay and the patient reports that the right side one (1.8) was lost six years ago by extraction. With no other associated pathology, the patient reported that she also suffered a surgery for a congenitally inguinal hernia during childhood, at eight years of age.

### Discussions

There are several studies in the literature that have attempted to determine the prevalence and type of missing teeth. Studies regarding the prevalence of hypodontia reported results ranging from 0.1% to 2.4% for primary dentition and from 0.15% to 16.2% for permanent dentition, excluding the third molars [24].



**Figure 8 – Panoramic radiograph of the aunt showing no other changes in the dental formula than the ones already mentioned by the patient.**

Another study, which analyzed the teeth absence, concluded that except the third molars, the prevalence of patients with congenitally missing teeth ranges between 1.6–9.6% [25]. Prevalence of missing teeth varies, depending on the population group. In a study conducted on the Iranian population, it was shown that the prevalence of congenitally missing teeth including the third molars is 45.7% and without the third molar, it is 10.9% [26]. A similar percentage of 11.2% was found in the population of Korea [27] and 11.3% in Slovenia [28]. In Germany, the results obtained regarding to the congenitally missing teeth prevalence showed a value of 12.6% [29], while the Australian and North American populations recorded lower values of 6.3% and 3.5%, respectively [30]. The prevalence of congenitally missing teeth was reported as being 2.6% for the South Arabian population and 11.3% for the Irish population [31]. A prevalence of 7.66% for congenitally hypodontia was reported in a study conducted on a Southern Iranian population [32]. Also, a different study in Iran reported a prevalence of 9.1% [33]. Another study shows that the prevalence and distribution of hypodontia in the pediatric orthodontic population in Venezuela is about 4% [34]. The percentages showing hypodontia prevalence in the general population worldwide are not high, showing that missing teeth are rare cases.

Regarding the type of tooth that is missing, it was observed that the last teeth belonging to each series are frequently involved in hypodontia, for example from the molars group, the third molar, from the premolars, the second premolar and from the incisors, the lateral incisor. In a study conducted in Iran [26], it was shown that the most frequent missing tooth is the mandibular second premolar, followed by the maxillary second premolar, the maxillary lateral incisors, and the maxillary first premolar. Another study conducted by Polder *et al.* [30] showed that the most common congenitally missing teeth are mandibular second premolars followed by maxillary first premolars and maxillary second premolars. Studies conducted on the Japanese [35] and Chinese [36] populations had similar results showing that the most frequent missing tooth is the mandibular second premolar, followed by the maxillary lateral incisor and the mandibular lateral incisor. Regarding the Korean population, it was shown that the most common congenitally missing tooth is the mandibular lateral incisor, followed by the second mandibular premolar and the maxillary second premolar [28]. The order of the frequency of missing teeth was

reported by Glenn [37] as the second mandibular premolars, the maxillary lateral incisors and the maxillary second premolars. The cases presented in this paper reveal the presence of hypodontia, probably congenitally, in second lower premolars, maxillary lateral incisors and mandibular central incisors.

It was analyzed whether congenital dental disorders are more common unilaterally or bilaterally. There are studies showing a unilaterally increased frequency of missing teeth [28, 30] and others showing a bilateral increased frequency [38]. This can be explained by the racial differences of the analyzed populations. In our study, the adult male case showed hypodontia situated both unilaterally and bilaterally, combined with size and shape variations, while both the female cases showed only unilateral hypodontia. The second patient has a unilateral hypodontia of right mandibular second premolar, while in the maxillary arch the two lateral incisors are smaller and conical; they are missing in her father's case. The father shows the bilateral absence of the upper lateral incisors and the bilateral absence of the central incisors. The aunt exhibits unilateral lower second premolar missing on the right side, the centrals and laterals both on the upper and lower arches are of normal form and dimensions. Cases in literature have shown an association between hypodontia and variations of the shape or size of the teeth in descendants; upper lateral incisors can present a reduced crown size and a modified shape, the mesial and distal surfaces of the tooth converging and forming a so-called peg-shaped crown [18, 19].

In order to early diagnose missing teeth, it is useful to have a panoramic X-ray after four years of age [39], showing the absence of permanent tooth buds. The panoramic radiograph at the age of nine years of the girl patient revealed the absence of the lower second premolar on the right and the absence of all third molars. Congenitally missing of all the third molars can be assumed; in her father's case, the maxillary third molars are present, while the inferior right one (4.8) is missing, and the inferior left one (3.8) has a smaller size and modified shape.

The challenge regarding the congenitally missing teeth is the therapeutic protocol. Some authors' recommendation of odontal treatment for isolated missing teeth is to keep the deciduous tooth on the arch for as long as possible and to protect it from occlusal abrasion and trauma [40]. Others suggest different treatment alternatives in case of missing lower premolar. A first option would be extracting the temporary molar and closing the space by orthodontic treatment [41]. Another treatment option would be to keep the temporary molar on the dental arch for a longer period of time and to perform a prosthetic or implant-prosthetic treatment when it will be lost [41–44]. If complications regarding the temporary molar occur, its resorption or ankylosis, the extraction and oral rehabilitation through other solutions becomes necessary. A dental implant could be inserted after the end of the growth of the jawbone. Jha & Jha [45] reported as a treatment method in the case of a 14 years old patient with bilateral missing inferior second premolars, cutting the deciduous mandibular second molars and retaining their distal halves, with coronary restorations in the shape of premolars. Other authors shown that, in cases of second premolar hypodontia, the deciduous second molar can last for a long

period on the arch [46]. Bjerklin & Bennet analyzed the persistence on the dental arches of the deciduous lower molars in patients aged approximately 30, and found that only two out of 59 teeth were lost, and this happened after the age of 20 [47]. In a similar study, Sletten *et al.* showed that out of 28 deciduous monitored molars, only four were lost and this happened at a mean age of 51 years [48]. Other studies also reported that the lifespan of temporary teeth may be increased [49–51]. In our case, as the deciduous mandibular right lower molar of the girl was not affected by decay and taking into account the results of the studies which demonstrate the great longevity of the temporary teeth on the arch, it was decided to retain the tooth in the oral cavity for as long as it will last, until adult age.

## Conclusions

Congenitally missing teeth are usually associated with other general conditions; still, in rare situations they can also occur as an isolate condition in clinically healthy patients. In our case, genetic transmission can be autosomal recessive or related to the sex chromosome. Thus, the transmission of right lower premolar missing from the aunt to the niece could be shown. The treatment of congenitally missing mandibular premolars is a challenge for dentists. The problem is the decision to extract the deciduous teeth or keep them on the dental arch. An early diagnosis of hypodontia is very important for preventing aesthetic, functional and psychological changes. Management of missing teeth treatment is complex, requiring close collaboration between specialists, such as orthodontist, prosthetician and implantologist.

## Conflict of interests

The authors declare that they have no conflict of interests.

## Authors' contribution

Authors #1 (RM) & #2 (OCA) have equal contributions to this paper.

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