Dental abnormalities in children with cleft lip with or without cleft palate

Agnieszka Lasota

Abstract

Introduction. Cleft lip with or without cleft palate (CLP) is associated with hypodontia, supernumerary teeth, dysmorphism of the crown or root shape, teeth impaction and malposition. The prevalence of anomalies is higher than in general population.

Objective. The aim of the review is description of dental anomalies affecting children with cleft lip or without cleft palate.

Abbreviated description of the state of knowledge. The most common anomaly is teeth agenesis with prevalence of 28% – 66% of patients with CLP. Permanent dentition is more often affected (52.7%) than primary (16.2%), the cleft-side lateral incisor is most often missing. Hypodontia increases with severity of the cleft. Supernumeraries were found in 17.7% of the subjects with cleft lip and/or palate for primary maxillary dentition, and in 5.7% for permanent maxillary dentition. Supernumerary teeth are mostly maxillary laterals, both inside and outside the cleft, and have different variations in morphology and size. The anterior maxilla is a main area for supernumeraries occurrence. Lateral incisor in the cleft area is often peg-shaped, its microdontia was revealed in 5.6%; they can also have enamel hypoplasia – 8.9% of cases. Macroodontia was found in 12.5% and taurodontism in 15.2%. Canine impaction prevalence is different in studies from various cleft centres and ranged from 0% – 58%. Rotations of central incisors was found in the area of cleft in 86.7%. Transposition of maxillary canine and first premolars was found in 5.5% of bilateral, 8% of right, and 3.3% of left unilateral clefts in the study.

Summary. Multiple teeth anomalies affect the population of children with cleft lip with or without palate. The most common problem is hypodontia. Treatment process of these patients demands experienced specialists in different fields of dentistry and complex, rigorous dental care.

Key words

agenesis, supernumerary tooth, cleft palate, dental anomaly, cleft lip

INTRODUCTION

Cleft lip with or without cleft palate (CLP) is the most common congenital face anomaly worldwide with the frequency 1:700 live births [1]. Over 80% of cases are non-syndromic clefts without congenital deformations. Cleft is associated with dental anomalies hypodontia, supernumerary teeth, dysmorphology of the crown or root shape, teeth impaction and tooth malposition. These anomalies are more frequent in permanent teeth, although they can also occur in primary dentition.

The etiology of dental anomalies in cleft patients was not fully recognized, but for some investigation it is believed that a genetic factor plays the main role. Among others, the gene candidates underlying the occurrence of clefts and congenital defects are MSX1, PAX9, IRF6 [2]. Howe et al. correlate the incidence of dental anomalies with local mechanical circumstances at the time of the cleft formation [3]. Absence of fusion between the maxillary and medial nasal processes that results in the CL/P, explains the problems affecting the lateral incisor and the presence of supernumerary teeth [4]. The influence of medical procedures is also emphasized.

Frequency of dental anomalies is higher in cleft patients than in the general population [5, 6], and the incidence of relative risk (IRR) of anomalies was highest in patients with bilateral CLP, followed by those with LCLP [7].

It is must be highlighted that children with oral clefts require interdisciplinary longitudinal treatment. Orthodontics is a main part of the process and is extremely difficult. Patients with CLP suffer from multiform skeletal malocclusions, usually class III with maxillary hypoplasia, class II, asymmetry and different types of vertical defects. Co-occurrence with dental anomalies impede treatment process because it is complicated, time-consuming and expensive. This indicates that the cleft anomaly is both a severe individual and social problem.

The review was carried out based on frequently cited studies from the PubMed and ScienceDirect databases.

OBJECTIVE

The aim of the review is a summarized description of dental anomalies affecting children with cleft lip with or without cleft palate.

TEETH ANOMALIES

Teeth agenesis. The main dental anomaly co-occurring with CLP is teeth agenesis, with a frequency higher than in the general population [8]. Congenital lack of teeth can be divided into three groups: 1) hypodontia – when one to five teeth are missing, 2) oligodontia – six or more teeth missing, and 3) anodontia – total lack of teeth. Hypodontia
was revealed in 28%–66% of the patients with CLP [3, 4, 9, 10]. Permanent dentition is more often affected (52.7%) than primary (16.2%). Hypodontia increased with the severity of the type of cleft [11]. Multiple hypodontia was found more frequently in the subjects with bilateral cleft lip and palate, and those with unilateral cleft lip and palate.[11]

It is interesting that the higher prevalence of agenesia, compared with non-affected subjects, exits even in the non-cleft area [12]. Jamalian et al. in their study did not reveal any significant difference between the genders in the prevalence of hypodontia, which differs from the healthy population because the female gender is prone to hypodontia [9].

Etiology analysis emphasizes the importance of the genetical factor. The molecular pathways, cellular functions, and tissue-specific expression of IRF6, explains the responsibility of this gene for 12% of genetic-influenced non-syndromic CLP, as well as proving IRF6 as candidate gene for dental agenesia [13, 14, 15]. IRF6 is responsible for the embryological formation of facial structures, including the teeth and tongue [16, 17, 18, 19]. This gene influences both epithelial adhesion during palate closing and odontogenetical oral epithelium differentiating [18, 20, 21].

An example of the co-occurrence of the CLP and teeth agenesia caused by the genetic factor is the Van der Woude syndrome (VWS; OMIM #119300) which, in 68 % of cases, is caused by IRF6 mutations or deletions [16]. VWS is characterized by paramedian lip pits and sinuses, conical elevations of the lower lip, cleft lip and/or cleft palate, and hypodontia with frequent absence of second premolars. There is a tendency towards greater maxillary hypoplasia, particularly in the most severe cleft type (bilateral CLP); the highest incidence of missing teeth is also seen in VWS with the more severe cleft type [22].

The tooth most often affected is lateral incisor on the cleft side, which was observed in 40% of the examined patients [23]. Location of hypodontia can be inside and outside the cleft region. When agenesia is found outside, this can suggest a genetic background, and usually affects the contralateral incisor, or less often, the second premolar in the maxilla or in the mandible. Other findings have revealed the second premolars as the most often affected. Studies concerning tooth agenesia has indicated that outside the cleft region, the prevalence of hypodontia is also increased in relation to healthy subjects [12, 24]. Mangione et al. describe the distribution in a large sample of individuals as being the same – inside (45.3%) and outside the cleft region (54.7%, of which 14.1% were in the mandible) [25]. The large amount of outside agenesia could result from the significant influence of MSX1 and TGFB3 genes in both hypodontia and orofacial clefting [26]. It is interesting that even in isolated soft tissue cleft, the agenesia and other dental abnormalities were more frequent on the affected side [27].

Korolenkova et al. found primary periosteoplasty and reduced blood supply associated with palatal defects as a reason of agenesia of maxillary central and lateral incisors [28]. On the contrary, the results of Tortora et al. indicated that early secondary gingiviovealveoloplasty has no influence on subsequent dental development [29].

Hypodontia in the cleft area is difficult clinically to solve. In growing patients, removable prosthetic devices or Maryland bridges are used, but they must be systematically adjusted to the growth changes. When fixed restoration is planned in adults, the lack of bone volume often enables implant placement, and the final result is often compromise.

**Supernumerary teeth.** Supernumerary teeth are defined as the presence of more teeth than the normal dental status [30]. They can be normal in shape or smaller in size, have an abnormal crown or root morphology; their developmental stage can also differ from the rest of the teeth. Supernumeraries were found in 17.7% of the subjects with cleft lip and/or palate for primary maxillary dentition, and in 5.7% for permanent maxillary dentition [11]. Pradhan et al. (2020, in an Iranian population of cleft children, revealed a 15% prevalence[31]. This is much more frequent than in the general population with a prevalence of 0.1% – 3.8% in permanent and 0.35%–0.6% in deciduous dentition. Supernumerary teeth may cause many clinical complications, such as in alveolar bone grafting.

Etiology of the anomaly has not been fully recognized. Fragmentation of the dental lamina during cleft formation as the cause of hyperdontia has been described [32]. Another hypothesis was lengthening of the precanine section of the oral epithelium caused by cleft, and thus an extension of the dental lamina which can develop into a supernumerary tooth. The division of the bud of the lateral incisor tooth d, situated across the clefted nasopalatal sulcus, could also probably lead to the formation of an extra tooth [33]. Non-fusion of the nasal and maxillary fields, or a potential post-fusion rupture of the cleft in the lateral incisor region, could produce the splitting of the tooth germ [25]. In the research by Scancelot on unilateral cleft lip and palate in children, the most commonly affected tooth was the cleft lateral [34]. In the study of Pradhan et al. (2020), supernumerary teeth were mostly the maxillary laterals, both inside and outside the cleft [31].

Variations in the morphology of supernumerary teeth involve normal shape and size, normal shape and reduced size, and conical shape. Single or multiple supernumerary teeth can be unilateral or bilateral, and can be located in the anterior maxilla and mandibular premolar regions. The clinical solution is usually extraction of the additional tooth, but sometimes there are difficulties in differentiating the normal tooth from an additional tooth. Cone-Beam Computed Tomography (CBCT) examination is obligatory for good clinical assessment and treatment planning in the case of supernumerary teeth. A limitation of the examination that must be considered is the lack of possible colour assessment of non-erupted teeth.

**Tooth shape and size anomalies.** In children with CLP, the upper incisors are often affected with shape anomalies. The lateral incisor in the cleft area is often peg-shaped or hypoplastic [23, 35, 36, 37]. Upper lateral incisor microdontia was revealed in 5.6% of the examined individuals with oral cleft [23]. Microformed lateral incisors were not found in primary dentition [11]. Lateral incisors can also have enamel hypoplasia which was detected in 18.9% of cases [23]. The etiology of this defect was described by Korolenkova as connected with primary periosteoplasty surgery and reduced blood supply [28].

In the study by Tan et al., 12.5% of examined patients had macrodontia, and all the cleft-sided permanent lateral incisors had associated anomalies [38]. An interesting finding is that posterior teeth can be bigger in size than in the healthy population [35], which suggests a multiple teeth size disorder in patients with cleft. Akcam et al. reported asymmetries in teeth dimensions comparing cleft and non-
cleft side [39]. Maxillary central and lateral incisors were larger on the non-cleft side in the mesio-distal dimension compared, with the cleft side. Upper central incisors and first molars are significantly larger mesiodistally on the non-cleft side [9].

Taurodontism was found in 15.2% of patients with CLP [40]. This is a molar tooth anomaly with an enlarged vertically crowned at the expense of the roots, and apically moved furcation. The probable mechanism of taurodontism is the failure or late invagination of Hertwig’s epithelial root sheath, and lack of shift of the root furcation.

Tooth shape and size anomalies cause asymmetry in the dental arch and poor esthetic appearance. It demands orthodontic alignment followed by prosthetic restoration of hypoplastic or deformed teeth.

**Teeth impaction.** Impaction of teeth is the lack of eruption after a time. Tooth impaction in patients with cleft can affect different teeth: upper incisors, canines and premolars. In the literature, canines impaction is described most often. The frequency of the anomaly is highly different in studies from varied cleft centres and ranges from 0% – 58%. An interesting finding is the higher incidence in small samples regarding bigger multicentre studies. An increased frequency of upper canines impaction in relation to the healthy population was described by Westerlund [41].

The etiological factor can be disturbed anatomy or genetic predispositions. The hypothesis is that the movement of tooth germs at the time of eruption leads to a disturbed position of those teeth in the dental arch, or to their complete impaction. Narrowed clefted maxilla and lack of space can also enable proper eruption. In the cleft are, a secondary bone grafting should improve eruption of an upper lateral incisor and canine; therefore, lack of exact treatment can be the reason orof for upper canines impaction. A Polish study by Pastuszak et al. did not confirm any relationship between single medical procedures; secondary bone grafting, maxillary expansion or extraction of non-resorbed primary canine, and prevalence of upper canine impaction [42]. The conclusion of the study was the importance of complex orthodontic care in improving of normal upper canine eruption in patients with CLP, emphasizing that initial position in bone of upper canine germ in most of cases is unfavourable.

In the general population there is a positive correlation between the occurrence of dental anomalies, such as hypodontia or reduced size of maxillary lateral incisors, and canines impaction, and this was not confirmed in the population with CLP [42].

Patients who suffer from tooth impaction demand surgical and orthodontic interdisciplinary procedure to achieve the tooth and align the dental arch.

**Teeth malposition.** In children with CLP, the maxillary teeth are more frequently in an abnormal position. A narrow and short upper arch means lack of space and consecutive crowding. Rotations, ectopic eruptions, transpositions are very common if the orthodontic treatment is not begun early. Early expansion and arch formation allows the avoidance of later complications. Unfortunately, some germs displacement are so severe that it is not possible to prevent malposition of erupting teeth. Rotations of central incisors is common problem in the area of the cleft with a prevalence of 86.7% [38]. A significantly greater frequency of rotations was found in the female gender [43]. Orthodontic derotation can be conducted if a sufficient amount of bone had been previously grafted. In cases with poor bone support, excessive movability of the central incisor can occur during orthodontic alignment. The initial CBCT examination allows assessment of the bone-root relation, planning of safe orthodontic treatment, or additional bone grafting before derotation.

Transposition of maxillary canine and first premolars was found in 5.5% of bilateral, 8% of right, and 3.3% of left unilateral clefts in the study of Eslami et al. [43]. Management depends on the level of transposition; in cases of total change of teeth position, a possible solution is leaving teeth transposed without intervention.

Common teeth malposition in CLP is palatal eruption of upper lateral incisors and upper second premolars due to lack of space and class III tendency.

All described teeth displacement demands comprehensive orthodontic fixed appliance therapy.

**CONCLUSIONS**

Multiple teeth anomalies affect the population of children with cleft lip with or without cleft palate. Congenital agenesis is the most common problem. It should be take into consideration that the treatment process of these patients demands experienced specialists in different fields of dentistry, and complex, rigorous dental care.

**REFERENCES**


