Journal of Dentistry & Oral Disorders

Open Access

Austin Publishing Group

Review Article

Hypohidrotic Ectodermal Dysplasia and Its Manifestations in the Oral Cavity

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Received: March 31, 2021; **Accepted:** April 28, 2021; **Published:** May 05, 2021

Abstract

The Ectodermal Dysplasias generally present orofacial manifestations, such as skeletal discrepancies and dental alterations. Therefore, the role of a paediatric dentist in the detection and recognition of these repercussions can be crucial in early diagnosis of the disease. The oral rehabilitation of paediatric patients with this condition is extremely important, ideally, at a very early stage, yet contributing for the re-establishment of normal chewing, swallowing and phonetics functions, and, naturally, aesthetics increase. The purpose of this narrative review aims to elucidate dentists about their role in the detection, diagnosis, treatment and monitoring of the Ectodermal Dysplasia' oral manifestations in paediatric patients, through the presentation of general physical and specific craniofacial characteristics.

Keywords: Ectodermal dysplasia; Oral manifestations; Dental anomalies; Paediatric dentistry; Hypodontia; Oral rehabilitation

Abbreviations

ED: Ectodermal Dysplasia; HED: Hypohidrotic Ectodermal Dysplasia; NFED: National Foundation for Ectodermal Dysplasia; OR: Oral Rehabilitation; VDO: Vertical Dimension of Occlusion

Introduction

Ectodermal Dysplasias (EDs) are a heterogeneous group of rare congenital diseases, which result from genetic disorders during the embryonic development phase in which, two or more structures of ectodermal origin are affected, with at least one of these changes occurring in hair, teeth, nails or sweat glands [1,2].

Distinct classifications have been attributed to EDs over time [3-6]. There are numerous types of known EDs [7] and the genetic mutations that give rise to them are numerous [3]. There is a wide spectrum of clinical presentations for individuals affected with this condition [8]. Among all the clinical manifestations of EDs, some appear, since very early, in the child's oral cavity, with several characteristic and representative signs and symptoms [9].

Dentists, especially paediatric dentists, due to the close contact established with children from an early age, have a privileged and extremely important role in the diagnosis and treatment of oral manifestations of this disease. In this sense, there is a need to know and recognize its possible orofacial manifestations [10], as well as the available and most appropriate clinical approaches for each child.

This narrative review aims to:

- Elucidate dentists about the importance of recognizing EDs and its orofacial manifestations.
- Emphasize the importance of preventive, therapeutic and monitoring dentistry measures, with the aim of improving the quality of life of EDs patients.
- Highlight the importance of a structured dental clinical approach adjusted to each child, defined by a multidisciplinary team, taking into account the different therapeutic modalities.

Considering the heterogeneity of the proposed classifications and the diversity of EDs typologies, for this work it was decided to approach the EDs in a general perspective and in a more in-depth way, its orofacial repercussions, concomitantly, with the various types of dental treatment currently available.

Ectodermal Dysplasia

In the third week of intrauterine life, the human embryo is already formed by three layers of cells, from which all tissues and organs of the fetus will be developed: ectoderm, mesoderm and endoderm. The outermost layer of the embryonic leaflet, called ectoderm, is responsible for the origin of several structures such as: the epidermis and its attachments (hair/hair and nails), the central and peripheral nervous system, the sensory epithelia of the sensory organs, the dental enamel and several glands, including sebaceous, sweat [4], lacrimal, mucous and salivary glands [11]. When genetic disorders occur at this stage of embryonic development, which result in an abnormal structure or function of ectodermal structures and, occasionally, structures derived from the mesoderm, a condition called Ectodermal Dysplasia is observed [12,13].

The term ED includes a diverse group of congenital disorders, which exhibit changes in at least two ectodermal appendages, with at least one of these changes occurring in hair, teeth, nails or sweat glands [2,14]. These four classic structures are affected in the following decreasing order of frequency: abnormal hair (hypotrichosis) (91% of cases), dental anomalies (80% of cases), dystrophic nails (onychodysplasia) (75% of cases) and structural and/or functional defects in the sweat glands (hypohidrosis or anhidrosis) (42% of cases) [2,15].

The inheritance patterns of EDs are variable, including all possible forms of Mendelian inheritance (autosomal dominant or

Citation: Cardoso JS, Faria Carvalho D, Carvalho Silva C, Moura Teles A, Leal F and Lopes Cardoso I. Hypohidrotic Ectodermal Dysplasia and Its Manifestations in the Oral Cavity. J Dent & Oral Disord. 2021; 7(2): 1161.

recessive), or linked to the X chromosome, also dominant (HD-X) or recessive (HR-X). In addition to these, the condition may also occur sporadically due to mutations during intrauterine life [3,16].

Several classifications of EDs have been proposed over time. Clouston, in 1939 [6], classified EDs in two forms, according to the degree of sweat gland dysfunction: hydrotic form (normal sweat production) and hypohydrotic or anhydrotic form (deficient or absent sweating, respectively). Later, in 1971 [4], Freire-Maia established a classification based on clinical criteria. Priolo et al. [5] suggested that EDs can be classified according to molecular genetic attributes; on the other hand, in 2003, Lamartine [3] proposed a classification based on the identification of ED causative genes. Recently, an international advisory group met at the National Institutes of Health in Bethesda to discuss a new classification system for the ectodermal dysplasias that would integrate both clinical and molecular information. The molecular causes of these diverse conditions involve many genes that code for enzymes of multiple developmental pathways and proteic components of complex molecular structures that are necessary for normal formation, structure, and function of the ectodermal derivatives [17].

EDs are heterogeneous both clinically and genetically. There are 163 well-established clinical entrances in Group A of ED. Mutations in seventy-seven genes have been proven to be responsible for 75 EDs [18].

The clinical characteristics associated with this congenital disorder can vary from case to case, depending on the type of manifested ED [19,20]. Hypohydrotic Ectodermal Dysplasia (HED) represents the most common phenotype of ED and is characterized by the presence of a triad of signs - hypotrichosis, hypodontia or anodontia and hypohidrosis or anhidrosis [21-23] - being, also, the most severe of all ED forms [23]. Frequently, it involves more severe dental anomalies [20]. The X linked form of HED, also known as Chris-Siemens Touriane Syndrome [24] is the most recurrent form of HED [25]. The diagnosis of EDs is based on observation of clinical manifestations [26] and genetic testing.

Orofacial manifestations

Patients with ED have a broad specific orofacial characteristic [28] which require special attention from health professionals. In addition to oral changes, pathognomonic facial features make these patients very similar to each other [10]. Besides, the usual complaint of these patients, either in childhood either in adolescence, is their concern about dental anomalies and facial appearance [29].

Craniofacial alterations: According to Saksena and Bixler [30], patients with ED present a generalized symmetrical reduction of the craniofacial complex. Clauss et al. [18] stated that dysmorphic craniofacial characteristics and bone structural anomalies are associated with the phenotype of ED patients, suggesting that the hypermineralization of craniofacial bones is also part of the clinical condition. Lesot et al. [31] suggested that osteoblastic and osteoclastic activity may be altered in these children, causing not only bone formation defects, but also an increase in jaws bone density.

Among the facial characteristics and deformities of the skull, these children may exhibit: reduced facial height, square and bulging forehead, frontal protuberances, prominent supraorbital ridges, pigmented and wrinkled eyelids with antimongoloid palpebral slant, prominent and pointed ears, short and wide nose with flattened bridge that gives a sad shape; hypoplastic nose alae, anteverted nostrils, receding or prominent zygomatic arches, depressed, hypoplastic and concave middle third of the face, large lip filter, protruding and thin upper lip, inverted lower lip, hypertrophic frenum linguae, and small, pointed chin with a deep groove [32]. The skin on the face may be thin, dry and smooth with hyperpigmentation of the periorbital and perioral areas and with fine, linear wrinkles [33]. The eyebrows hair and eyelashes may be sparse, poorly developed, or absent [24,34]. Other characteristics such as the underdevelopment of the zygomatic bone and subsequently the resection of the zygomatic region [35], decreased maxillary and mandibular growth, deficient development of the maxillary and mandibular alveolar ridges [36] and hypotonicity of the perioral muscles [37] can also be observed.

The poor development of the jaws resulting from hypodontia leads to a decrease in the amount of alveolar bone in height and width, resulting in craniofacial skeletal discrepancies present in these patients [38]. However, it is important to note that the apparent skeletal changes are less in the mandible when compared with the maxilla [39]. In a retrospective study involving six hospital-based cases, it was found, according to cephalometric analyses, that the dento-maxillofacial characteristics such as hypoplasia and maxillary retrusion, counter-clockwise rotation and mandibular protrusion, were common among EDs patients [40]. In a similar study, it was found that in 8 of 15 cases studied through Steiner's cephalometric analyses, the individuals had a skeletal class III profile, associated with maxillary retrusion and mandibular protrusion, mentioning that these cases are more evident in clinical cases of extensive hypodontia [41]. Some authors report the presence of a reduced vertical dimension of occlusion (VDO) resulting from the reduction in the average growth of the face and the insufficient height of the alveolar edges due to hypodontia, that leads to a decrease in the lower facial height [6,29]. Craniofacial deviations tend to worsen with advancing age in untreated patients [42,43], and in addition to the unfavourable aesthetic appearance, they present decreased masticatory forces, a lack of functional occlusion and subsequent speech problems [44]. When these characteristics are manifested at the same time in a child, their facial appearance is similar to an elderly person [35,45].

Oral disorders: These patients often show xerostomia resulting from hyposalivation caused by the decrease in the number and the salivary glands' secretion, which are of ectodermal source [11,46]. In 2010, Bergendal [47] reported that 30.2% of these patients had reduced salivary flow rates. This condition is associated to difficulties in ingestion and chewing food, lack of taste and smell, as well as dysphagia and dysphonia [47,48]. In addition, hyposalivation can be considered a predisposing factor to the development of dental caries [11,49], as well as other infections at the periodontal tissues. Other oral manifestations may also include reduced vestibular depth, flat palate shape [32], underdevelopment of tuberosities and alveolar edges, atrophic gums [48] and soft tissue retrusion as a result of maxillary retrusion [33].

Dental anomalies: Dental anomalies are observed in 79% of patients with ED [50] and can affect both primary and permanent dentition [32]. These dental conditions can have multiple phenotypes [18,42], ranging from dental agenesis, size and shape variations,

mineralized tissue defects, as well as problems in tooth eruption [51].

Tooth eruption: Delay in tooth eruption is a common oral feature in these patients [52,53]. The order of dental eruption is often different from the common eruption chronology [29]. According to Wright et al. [54] the EDs diagnosis is more frequent at the age 6-9 months of the child, as it corresponds to the expected period of the first teeth eruption. If not erupted, or, when present, but with morphological changes, this should be a strong signal to tigger an accurate study of the cause. Including, the National Foundation for Ectodermal Dysplasias (NFED) recommends that if a child does not have any erupted teeth by the age of one, the suspicious of ED condition should be evaluated [55]. Other changes, such as the prolonged retention of primary teeth that can cause ankylosis can also be observed in the patients [10].

Dental anomalies of number: The absence of primary and/or permanent teeth is the oral characteristic most striking and frequently described in individuals with ED [52]. According to the number of missing teeth, agenesis can be classified into hypodontia, oligodontia and anodontia [57,58]. In fact, hypodontia can be considered a universal characteristic [56], frequent in 80% among these individuals [15]. Variations in the degree and location of agenesis in the dental phenotype can be caused by epigenetic factors [59,60], or by differences in the penetration of the mutation [61]. Generally, there is a higher frequency of dental agenesis in the lower jaw [24]. A study conducted by Dhamo et al. [62] revealed that the most absent teeth were the central incisors, second premolars, second molars and lateral incisors, while the first upper molars and lower canines were the least absent. Dental agenesis and their side effects on the growth and development of the jaws are predominantly the most significant clinical manifestation [63]. Maxillofacial growth is affected by the absence of teeth. In edentulous areas there is a weak development of the alveolar edges and, consequently, a reduced volume of the alveolar bone [64], which presents a thinner shape in contrast to the alveolus that surrounds the present teeth [56]. As bone development follow the formation of dental germs, when they are absent, bone formation is found to be dysplastic or abnormal [55]. This situation will result in a decrease in the bone volume available for oral rehabilitation (OR) with conventional prostheses or implants [65]. In addition, reduced bone growth caused by dental agenesis can induce bone deformation and teeth support problems due to distortion in the bone resorption-apposition process [32]. In addition to dental agenesis, supernumerary teeth can also be observed [10].

Dental anomalies of size and shape: Both primary and permanent teeth are frequently present with shape deviations, such as small size (microdontia) and widely spaced [66]. Incisors and canines are most affected by the conical and pointed shape, often described as "peg-shaped" [67]. Moderate to severe taurodontism is frequently observed in molars [68], mainly in deciduous lower second molars [18]. This can occur because the Hertwing' epithelial sheath that determines the pulp soil level and subsequentl the root bifurcation site is of ectodermal origin [55]. The small size of the roots [69], as well as the presence of posterior pyramidal and fused roots [67] can also be observed.

Dental anomalies of structure: Hypoplastic teeth are often described in patients with ED [67]. The thickness of teeth enamel in these patients is thinner than usual, especially in the occlusal and incisal regions. Dhamo et al. [62] observed a general trend in the delayed maturation of all permanent teeth in this condition. Dental crown shape is determined by the way the enamel is deposited on the dentin layer. Since enamel is the only dental tissue of ectodermal origin, it becomes the main dental structure related to EDs. Therefore, the abnormal formation and mineralization of the enamel can influence crowns shape and modify the developmental stages of the tooth due to the calcification process [62]. The enamel can be hypoplastic, hypocalcified or hypomatured, predisposing the individual to the development of caries and the appearance of changes in tooth colour, concomitantly with the high risk of tooth structure loss due to weak resistance to fracture [55].

Role of the dentist in the approach of the child with ectodermal dysplasia

The paediatric dentist has an extremely important role in the diagnosis of ED because he can often be the first health professional to be sought by parents. Therefore, it becomes necessary and crucial for this professional to recognize the possible changes inherent to this condition [10], perform a good anamnesis and take complementary diagnostic exams whenever necessary [67]. Once the diagnosis is established, an appropriate treatment plan must be developed, in accordance with the child's needs, listing the possible clinical approaches, and explaining to the child guardian the benefits and complications associated with each [70]. However, addressing all orofacial manifestations resulting from this condition requires a multidisciplinary team, which should include a paediatric dentist and other health professionals with clinical skills in other medical fields, to ensure an appropriate treatment and achieve the best functional and aesthetic results [20,71,72]. The treatment of a paediatric ED patient requires knowledge and experience about child growth and development, behavioural control, the ability to motivate the child and parents to the use of prostheses or the placement of implants and to do the medium and long period follow-ups [73].

Preventive and therapeutic approaches for children with ectodermal dysplasia

There is no specific dental treatment for children with ED [74]. These patients should receive multidisciplinary dental care at an early stage to minimize the impact on their quality of life [8,75]. Rehabilitation of these patients should help to restore their chewing and skeletal growth patterns, and reduce the consequences associated with the loss of VDO and the development of malocclusions [53].

Dental treatment must be tailored, including therapeutic options aimed at the specific needs of each patient, with the main goal of orofacial development and the restoration of function (chewing, swallowing and phonetics) [76,77]. After the diagnosis of this condition, all possible approaches should be presented to the child and their guardians, which may include preventive measures for the control of the main oral diseases, assessing risk factors, such as, oral hygiene and diet habits [75]. On the other hand, it can include therapeutic measures such as restorative, periodontal, endodontic, prosthodontic, orthodontic and surgical treatments [8] and their lifelong monitoring. Rehabilitation treatment options can range from direct or indirect restorations with composite resin [78], to ceramic or metal-ceramic crowns or bridges [63], partial or total prostheses,

fixed or removable, implant prostheses [79] and overdentures [80,81].

These therapeutic approaches can be used individually or combined to obtain the best results [79,82]. The chosen rehabilitation option must allow a correct growth pattern for the child [8,29]. Although there is no ideal time to start the OR of these children, several authors state that rehabilitation should start before attending school for the first time, due to facial aesthetic and its influence on the development of interpersonal relationships and social acceptance [79,83]. In children with anodontia or severe oligodontia, early prosthetic treatment at less than five years of age is reported in most cases and seems to be favourable in terms of food intake, development of speech, and socialization [84]. In a study of Ding et al. [35], the quantitative analysis of the cheeks volume of ED patients revealed that rehabilitation has a fundamental role in restoring facial support, since the cheeks are sunken in these patients. The same study also showed an improvement in facial appearance, at the level of the lips, zygomatic region and chin, after the rehabilitation of lost teeth thus creating support for the facial musculature and consequently facial fullness [35].

Before starting any rehabilitation treatment, it is important to motivate and raise awareness of the child and guardians about the importance of oral hygiene and prosthetic care [85], and the need of frequent dental appointments to allow the necessary prosthetic adjustments related to the child's continuous growth and development [79,86-88]. Oral hygiene and dietary instructions, concomitantly to the teeth cleaning and topical application of fluorides should be part of the monitoring appointments, since these children are at high risk for dental caries development due to hyposalivation [33,41] and enamel dental defects [55]. The dental materials used in the OR of these patients must allow periodic modifications due to maxillary development besides dental eruption process [63].

Prior to planning these children's OR, the following criteria should be considered: the patient's age, the craniofacial growth and development, the available alveolar bone volume, the degree of hypodontia, the existing intermaxillary relationship, the dental anomalies of number and position, the presence of dental germs [38,65,89-93], the masticatory and phonetic functional state, the aesthetic, emotional and psychological aspects of the child [94,95], the motivation of the patient and the family economic resources to support the costs of treatment [48,88].

Discussion

A wide variety of dental treatments, especially restorative ones, are suitable for ED children. However, it is important to emphasize that whenever possible the preservation of primary and permanent teeth is desirable in preventing loss of alveolar bone required for support in prosthetic retention and in orthodontic anchorage [94,96].

There are many options for OR for ED patients [91]. Several authors state that direct or indirect restorations with adhesive composite resins are an adequate, non-invasive solution for conservative, aesthetic and reversible restoration of teeth having changes in shape, size and structure [78]. This approach is often described in the literature combined with the use of removable partial dentures [87].

Conventional removable protheses are the dental treatment frequently reported for the OR of children with hypodontia or anodontia [28,48,97,98]. Several types of prosthetic rehabilitation with removable prostheses are suggested in the literature [98]. However, although these prostheses can provide an acceptable aesthetic and functional result, several authors report that ED is a difficult condition to treat with this rehabilitation approach due to the patient's young age and their orofacial changes [83], such as: abnormal dryness of the mucosa (which acts as potential cause of prothese' rejection as it can easy damage the soft tissues), underdevelopment of tuberosities and alveolar edges [99] and the presence of conical and poorly positioned teeth that hinder their retention and stability [100]. Other authors, despite considering this option as "the gold standard", they highlight that it can lead to complications, such as increased number of caries and periodontal complications if due to not improved oral hygiene tacit to protheses' use [70]. The prostheses should allow for periodic modifications as maxillofacial growth and tooth eruption occur. Being so, metallic components should be avoided in the design of the prosthesis [63] giving preference to acrylic prostheses options that more easily allow adjustments [91].

With regard to fixed prosthetic rehabilitation, rigid connectors should be avoided in patients in the growth phase, specifically if the prosthesis crosses the dental midline, as they can interfere with the normal jaws' development [79].

The OR of paediatric patients with implant-supported prostheses has been frequently reported as an alternative to conventional removable prostheses due to their better aesthetic and functional results [92]. However, OR using implants in these conditions generates some controversy among authors since the minimum age for implant placement is also not consensual. The orofacial changes that result from growth during childhood are not favourable to the maintenance of implants [101] due to their behaviour similar to ankylosed teeth, unable to follow craniofacial skeletal growth [102,103] resulting in functional and aesthetic disadvantages [39]. In this way, when implants are placed adjacent to natural teeth, jaw growth and the continuous eruption of surrounding teeth will lead to submersion and infra-occlusion of the implant [97,104]. In addition, when implants are placed in the growth phase, they may lead to a higher probability of peri-implantitis and fracture, since in these conditions, longer abutments are usually required and consequently, a peri-implant gingival groove deeper than normal [105]. According to Rossi et al., the placement of implants in childhood can interrupt the dental germs development and tooth eruption resulting in severe trauma to the patient [103].

In opposition, Guckes et al. [65] suggested that paediatric ED patients with extensive hypodontia or anodontia may benefit from an implant-supported OR during childhood, since treatment with conventional dentures presents retention, stability and support problems. According to Kramer et al. [94], the most suitable place for implants is the anterior region of the lower jaw because in this region, changes in growth are minimal after 7 years of age. Since the mandibular symphyseal suture usually closes at around 6 years of age, it is unlikely that after that, an increase in the transverse width of the anterior mandible occurs and consequently, changes in implants position would be minimal. In contrast, placement

in the upper jaw should be avoided until adulthood, or ultimately, prosthetic rehabilitation should not cross the midline due to continuous transverse growth [93]. Kramer et al. [94] consider that it is very difficult to determine the optimal period for implant placement, since several aspects must be considered simultaneously: the degree of skeletal growth, the degree of hypodontia, the condition and size of the teeth and the extent of psychosocial implications and stress of each patient. However, Thilander et al. [104] stated that the most important factor in determining the ideal period for OR with implants is the degree of skeletal and dental maturation, and not the age of the patient.

Klineberg et al. [106] suggested that anodontic patients aged between 5 and 10 years old may benefit from the use of prostheses on implants, when applied to the mandibular canine region and in the maxilla in children from 6 to 10 years old. The same authors found no consensus regarding the OR with implants from children aged 6 to 10 years with hypodontia and claim that implants should not be placed adjacent to the teeth until growth is complete. According to Sharma and Vargervik [94], placing implants in children can be balanced according to three different situations: children with agenesis of an isolated tooth should not be submitted to implant placement until growth is complete; anodontic children can benefit from this strategy from 7 years of age; and finally, children with multiple agenesis should be treated with removable prostheses until the end of growth, after orthodontic correction, as long as it allows satisfactory aesthetic and functional results. On the other hand, the NFED treatment guidelines indicate that growing children should benefit from an OR with non-rigid removable or fixed prostheses and that rehabilitation using implant prostheses may be indicated for school-age children (7-12 years old) and adolescents (13-18 years old) in the anterior region of the lower jaw, being contraindicated in the posterior mandibular regions and in the entire maxillary arch, since at these ages considerable growth is expected in these regions [55]. The Delphi study recommends placing implants in the anterior region of the mandible from 7-8 years old while in the maxilla only after the completion of growth [106].

Bergendal et al. [90] consider that the main risk factors for osteointegration of implants in ED children are: low amount of available bone, small mandibular dimensions, dense cortical bone and "loose" spongy bone. In addition, the higher bone density of the jaws is seen as a concern in the osteointegration of implants since the risk of bone overheating is higher [107]. Other researchers consider mini-implants as a viable provisional solution in these children's OR before they finish their growth, due to advantages such as the reduction of the implant diameter, shorter surgical protocol, absence of submerged healing, immediate prosthetics restoration and lower costs [28,108].

Although OR with implants of ED patients is often described with high success and survival rates [65,109,110], some authors claim that long-term clinical studies are necessary in order to reach consistent conclusions [111]. According to Mishra et al. [70], the use of this therapeutic option is only justified when the expected positive effects are greater than the disadvantages that may result from it. Therefore, if the child's oral health favours OR with implants, before completing its growth, their guardians should be informed about all the benefits and possible complications of its use. Oesterle et al. [112] recommend that, in the first rehabilitation phase, the paediatric patient undergoes the use of a removable conventional prosthesis, to gather aesthetic information required in the design of a future final prosthesis, and to allow the greatest possible growth before starting a treatment involving implants.

For some authors, the dental treatment of young patients using overdentures may be a conservative, simple and cheap alternative [80,113], when teeth' roots with a good support prognosis are present [79]. This option, besides allowing adjustments to craniofacial growth, has other advantages when compared to conventional full dentures: greater support and stability of the prosthesis, better mechanism of proprioception and neuromuscular feedback, function comfort, greater sense of security and preservation of the alveolar bone [80,113]. In addition, the presence of conical shaped teeth can be advantageous when used as abutments for overdenture [114]. Regardless of age, removable full or partial prothesis, or overdentures present a frequently applied treatment modality in children [84]. According to the NFED, small tooth movements, endodontic treatments and selective extractions may be necessary to enable successful prosthetic rehabilitation in these patients, from early childhood to adulthood [55]. Orthodontic fixed or removable treatment is often included as part of the medical/dental treatment, since it allows the modification of the pattern of dentofacial development and the position of the teeth, to create spaces that may be useful for prostheses or implants and allows the control of dental eruption and diastemas closure [53,64,94].

ED patients with severe skeletal deformities may need surgical correction to create favourable conditions for OR. Orthognathic surgery is generally used for jaws reposition, facilitating rehabilitation treatment and improving skeletal relationships and facial aesthetics [111,115,116]. Other surgical procedures such as bone grafts and elevation of the sinus membrane may be necessary and considered before rehabilitation with implants due to the severe bone atrophies observed in these patients [93]. Nevertheless, whenever possible these surgical interventions should be postponed until the craniofacial growth is complete [55].

Follow-up appointments should be scheduled considering several factors such as the child's age, the oral rehabilitation performed and the growth and individual development patterns. Specifically, prosthetic overdenture should be performed every 2-4 years and replacement of prosthesis every 4-6 years in a growing patient [81].

Conclusion

The dentist, especially the paediatric dentist, plays a particularly important role with regard to the early diagnosis of this condition, through the recognition of the orofacial manifestations that children with ED can present from an early age. It is often seen as the first health professional to be sought out by these children, due to oral changes frequently manifested, such as the absence of tooth eruption. However, he must be part of the multidisciplinary team, being able to diagnose, monitor and implement all the necessary dental care in the different life stages of the ED child, to improve his oral health and quality of life. Early medical/dental interventions will allow to maintain, correct or restore oral functions and improve facial aspect.

In addition, the sooner the growth anomalies caused by edentulism are intercepted, the smaller the extent and the easier the children's rehabilitation will be. The treatment plan must be carried out carefully and individually adjusted to the characteristics of each child. The OR of these children can be performed using conventional removable prostheses, prosthetic or implant-supported prostheses, taking into account several factors such as the child's growth and development, the degree of edentulism, the available bone support, intermaxillary relationship and the motivation of the child and parents for treatment. Orthodontic and/or surgical treatments may also be necessary. In addition, it is relevant to emphasize the importance of periodic and lifelong monitoring. The choice of treatment plan for these children must be cautious and its benefits must outweigh possible losses.

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