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Oral findings in pediatric patients with ectodermal dysplasia: A systematic review

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Abstract
Background: Ectodermal dysplasia (ED) is a rare group of hereditary disorders affecting tissues derived from the ectoderm, which includes teeth, hair, nails, and sweat glands. Many pediatric patients with ED often present with notable oral and maxillofacial abnormalities, such as hypodontia, oligodontia, abnormal tooth morphology, and craniofacial deformities. Despite this rising interest in these manifestations, a systematic synthesis of the evidence regarding their prevalence, characteristics, and clinical implications is lacking.
Methods: This systematic review was carried out in accordance with PRISMA guidelines. The search was performed in six databases by applying Boolean operators and MeSH terms. Oral and maxillofacial findings in children diagnosed with ED were considered for the study, which covered case series and observational studies. The data were extracted regarding demographic characteristics, dental phenotypes, craniofacial abnormalities, and treatment outcomes, which were analyzed. The ROBINS-I tool was used to assess the bias.
Results: A total of eight studies was included, with patients from different geographic locations and subtypes of ED. Dental findings included hypodontia (82.4%), oligodontia (61.8%), and anodontia (37.6%). Morphological anomalies like conical teeth (67.2%), paurodontids (29.3%), and enamel hypoplasia (48.1%) were present frequently. Craniofacial features included reduced alveolar bone height (54.6%), vertical facial deficits (43.7%), and narrow palates (38.5%). Salivary dysfunction was present in 26.8% of the cases. Radiographic assessments showed widespread dental agenesis and delayed eruption in more than 80% of the patients. Improved functional and aesthetic outcomes were associated with early prosthetic and orthodontic rehabilitation.
Conclusion: These findings reemphasize the heterogeneity of oral and craniofacial manifestations in children with ED. There were large variations seen among subtypes and population groups. In many children, early intervention with an individually tailored approach could prevent most functional and psychosocial impairment. These findings will offer a basis for enhanced diagnostic and therapeutic strategies in handling ED.

Keywords: Ectodermal dysplasia, pediatric patients, oral findings, hypodontia, craniofacial anomalies, prosthetic rehabilitation

Introduction
Ectodermal dysplasia (ED) refers to a heterogeneous group of inherited disorders characterized by anomalies in tissues derived from the ectodermal germ layer [1]. These conditions include both syndromic and nonsyndromic phenotypes and varying degrees of involvement of structures such as hair, nails, sweat glands, and teeth. Among the most clinically significant oral manifestations are hypodontia, oligodontia, anodontia, delayed eruption of teeth, and abnormalities in tooth morphology, including conical or peg-shaped teeth [2-3]. These dental anomalies have a profound effect on the function and aesthetics of the oral cavity and thus pose considerable challenges to the management of affected pediatric patients.
The most common subtype is hypohidrotic ED, usually inherited in an X-linked recessive pattern and with the males being more severely affected. In this subtype, the oral cavity is severely affected, leading to functional impairments in mastication, phonation, and overall oral health [4]. Accompanying these dental defects are associated abnormalities in the maxillofacial structures, including reduced alveolar bone height, compromised vertical dimension, and hypoplasia of the midface [5]. These often have interdisciplinary care by pediatric dentists,

orthodontists, prosthodontists, and oral surgeons.

The pathogenesis of ED is related to the mutations of several critical genes such as EDA, EDAR, and WNT10A that disrupt epithelial-mesenchymal signalling pathways during embryonic development [6-8]. These disruptions lead to abnormalities in tooth bud formation and differentiation, thereby contributing to dental defects that are noted in the disorder. The degree of variability in phenotypes observed amongst ED subtypes underscores the spectrum of molecular and clinical conditions noted in this condition [9]. Such knowledge is quite important for the diagnosis and prognosis of this condition; it will also allow precise tailoring of management according to the individual needs of pediatric patients.

The immediate physical consequences of the oral manifestations of ED extend far into the clinical arena. Children with ED, therefore, tend to be psychologically disturbed because their dental anomalies are very noticeable and may contribute to decreased self-esteem and social problems [10]. Functional deficiencies such as defective mastication may lead to nutritional deficiencies and disrupted growth patterns. Such difficulties, therefore, present a case for early intervention and comprehensive management in order to prevent or minimize the effects of ED on the child's well-being [11].

Despite the advances in genetic and clinical research, there is a lack of systematically reviewed data on the oral findings associated with ED in pediatric populations. The literature is dispersed, and studies vary significantly in methodology, sample size, and focus. The variability makes it essential to have a comprehensive synthesis of the available evidence to give a more coherent view of the oral manifestations in pediatric ED patients and their implications for clinical care. Hence, this systematic review tries to make an exhaustive report on the oral findings of children affected by ED regarding prevalence, morphological features, and associated maxillofacial anomalies.

Materials and Methods

PECOS Protocol Design and PRISMA Consistency

The PECOS protocol for this systematic review was carefully designed to ensure proper and unbiased identification of studies that would be relevant. This was strictly in compliance with the reporting guidelines in PRISMA 2020 [12]. The inclusion criteria were defined in the protocol as follows: Population (P), children patients diagnosed with any type of ED; Exposure (E), existence of ED focusing on oral and maxillofacial findings; Comparators (C), as appropriate, included the pediatric populations without ED or other syndromic conditions; Outcomes (O), including in-depth descriptions of dental anomalies (such as hypodontia, anodontia, enamel hypoplasia), maxillofacial characteristics (such as alveolar bone height), and salivary gland function; and Study design (S), including case series, cohort studies, and clinical studies. The protocol was designed to systematically select studies over a broad range of time and space so that data synthesis would be comprehensive.

Inclusion and Exclusion Criteria

Studies on pediatric patients, aged ≤18 years, diagnosed with any type of ED were included in the criteria. The included studies had a detailed description of oral and maxillofacial findings that included dental anomalies, morphological features of the teeth, and assessment of salivary glands. Only primary articles written in the English language were considered; case series, cohort studies, and clinical studies. Studies with clear focus of oral health outcomes in patients with ED were considered first and foremost.

Studies with an adult population or those where the focus is not defined on pediatric populations were ruled out. Articles that described oral findings without detailed analysis, review articles, abstracts without full texts were excluded. Editorials, commentaries, and review articles were excluded. Moreover, studies with a concentration on genetic analysis or simply unrelated systemic manifestations of the disease were excluded for maintaining clear scope.

Database Search Protocol

The database search protocol was developed to comprehensively identify relevant studies across six databases: PubMed, Scopus, Web of Science, Embase, Cochrane Library, and Google Scholar. The search strategy used Boolean operators and MeSH keywords. The primary search string was: ("ectodermal dysplasia"[MeSH] OR "hypohidrotic ectodermal dysplasia" OR "anhidrotic ectodermal dysplasia") AND ("oral findings"[MeSH] OR "dental anomalies" OR "tooth agenesis" OR "hypodontia" OR "anodontia" OR "enamel hypoplasia") AND ("pediatric"[MeSH] OR "children" OR "adolescents"). Filters were applied to limit the results to English-language studies and to studies involving human subjects. The search was customized for each database, using specific indexing terms where appropriate.

Data Extraction Protocol and Selected Data Items

The data extraction protocol was designed to systematically capture relevant information from each included study. A structured data extraction form was developed and piloted to ensure consistency. Data items extracted included identifiers (author name, year, location), study design, sample size, age, and male-to-female ratio. Detailed information obtained from oral findings included dental anomalies prevalence and distribution; existing teeth morphological characters, maxillofacial parameters, salivary gland assessments, periodontal health indicators, and radiographic findings. Additional items of data consisted of the genetic subtype of ED, when known, and any appropriate inferences to clinical effects or response to treatment. Data extracted were cross-checked by two independent investigators to minimize error and omissions.

Bias Assessment Protocol

Risk of bias of studies included was assessed with ROBINS-I tool: Risk of Bias in Non-randomized Studies - of Interventions [13]. This tool was used to assess bias across seven domains, namely confounding, participant selection, intervention classification, deviations from intended interventions, missing data, measurement of outcomes, and selective reporting. Every domain was assessed as a low, moderate, serious, or critical risk of bias. Conflicting bias judgments were settled by consensus of two independent reviewers to achieve unbiased judgment.

Results

Total 348 records were identified through database searches with no further records from the registers. From 29 duplicates removed, the remaining 319 records were screened through titles and abstracts with no records being excluded at this stage. Of the 319 records that were searched for, 44 could not be retrieved due to unavailability. A total of 275 records remained eligible for the full-text review. Of these, 197 records were excluded, including 76 literature reviews, 57 in-vitro studies, 62 animal studies, and 72 studies targeting adult populations. Finally, eight studies [14-21] were included in the review for qualitative synthesis, with no new reports added subsequently.

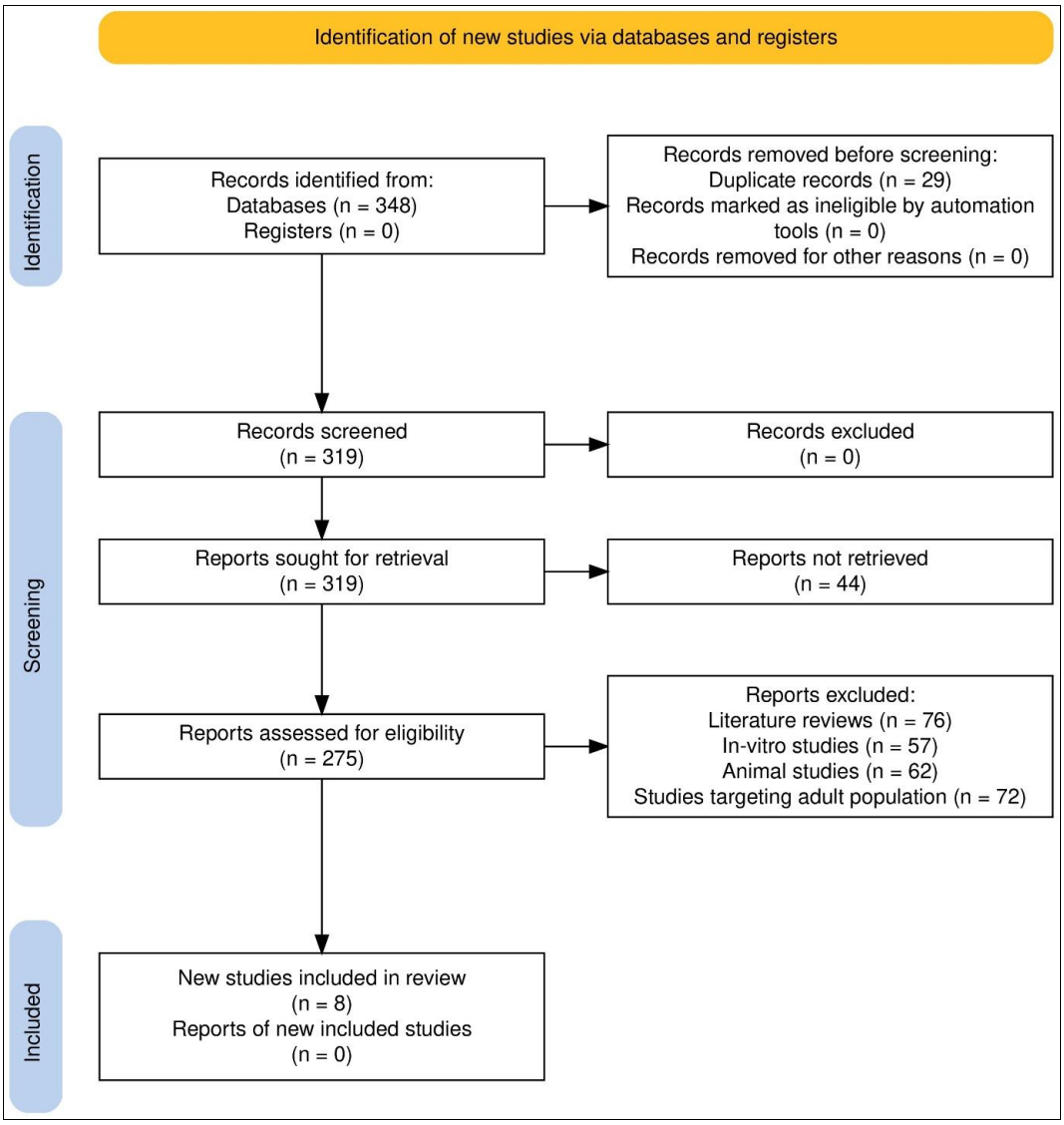

Fig 1: Study selection process representation for the review

Demographic Variables

The studies collectively analyzed ED in various geographic regions: Italy [14], India [15, 20], Turkey [16], Germany [17, 18], Greece [19], and Japan [21]. This broad geographical scope would ensure that the findings were population-specific and varied across different regions with regards to phenotypic expressions of ED (Table 1).

The studies included a duration from 2006 [18] to 2023 [15, 17] and signify the interest in exploring oral presentation of ED within nearly two decades. Sample size varied as it ranged between three [19, 20] patients and very large sample size of 1067 [17] providing insights between single cases and larger population-level studies. The mean age of participants ranged from young children (3.5 years [19]) to adolescents (15-19 years [20]), ensuring representation across critical developmental stages.

Gender distribution also differed, with male predominance noted in studies reporting severe hypohidrotic phenotypes [20], whereas other studies reported near-equal distributions [14, 21]. Follow-up durations ranged from short intervals (2 years [19]) to longitudinal assessments spanning 8 years [14], reflecting variability in study designs and their capacity to capture developmental changes over time.

Table 1: Demographic Variables Elucidation

Author ID	Year	Location	Study Design	Sample Size	Mean Age (Years)	Male: Female Ratio	Follow-Up Period
Dellavia et al. [14]	2010	Italy	Longitudinal Study	12 patients	8-15 (multiple intervals)	6:6	8 years
Goswami et al. [15]	2023	India	Case Series	4 cases	4-13	3:1	Regular follow-ups
Gürcan et al. [16]	2020	Turkey	Case Series	22 patients	10.54 ± 4.59	13:9	NA
Nadolinski et al. [17]	2023	Germany	Retrospective Study	1067 patients	4.5-18	594:473	6 years
Präger et al. [18]	2006	Germany	Clinical Study	30 patients	7-23 (mean: 12.3)	19:11	NA
Seremidi et al. [19]	2022	Greece	Case Series	3 patients	3.5 years	3:0	2 years
Sushitha et al. [20]	2022	India	Case Series	3 patients	15-19 (approx.)	1:2	NA
Terada et al. [21]	2012	Japan	Clinical Study	4 patients	7-12	4:0	NA

Oral Findings and Assessments

The studies described varied types of ED, including HED [14, 15, 20, 21] and mixed types [16, 17], so that ED seemed to exhibit clinical variation broadly. There was more consistent evidence for mutations affecting some of the most important key regulatory genes including EDA, EDAR, and PAX9 in many patients [14, 17, 20], underlining epithelial-mesenchymal disrupted signalling pathways in causing ED (Table 2).

Hypodontia, oligodontia, and anodontia were the notable dental phenotypes with subtypes of ED showing variable manifestations. The lateral incisors and molars were usually absent, while conical teeth were often present according to multiple reports [14, 16, 18]. Severe conditions usually involved anodontia in either the primary or permanent dentitions [14, 21]. Morphological anomalies with enamel hypoplasia, taurodontism, and malformed molars had recurrent occurrences [15, 16, 18] and emphasize the extent of developmental disturbances due to ED.

Maxillofacial features were typically characterized by reduced alveolar bone height, vertical facial dimension deficits, and narrow palates [14, 16, 21]. Such findings reflect the functional and aesthetic burdens of craniofacial deformities in ED. Dysfunction of salivary glands, when noted in some reports, varied from decreased salivary flow [15, 19] to normal gland function in milder cases [14], indicating variable systemic impact.

Less commonly reported were the periodontal and mucosal health evaluations; however, in those reported, good periodontal health was mostly observed in all cases [16], whereas mucosal dryness was associated with decreased salivary flow [19]. Radiographic findings presented universally with extensive dental agenesis and delayed eruption patterns [14, 16, 18, 21]. With advanced imaging modalities, such as CBCT and 3D scans, further maxillofacial anomalies were noted, such as hypoplastic alveolar ridges and diminished bone growth [14, 21].

Prosthetic and orthodontic rehabilitation approaches were a central theme in the majority of the studies. Interventions involved gap closures, removable prostheses, and orthodontic appliances that were intended to enhance mastication, speech, and aesthetics [15, 17, 19]. Early rehabilitation was strongly recommended to foster functional growth curves and reduce psychological effects of ED-related anomalies [14, 21].

Bias Levels Observed

The ROBINS-I tool was used to systematically evaluate the bias assessment across the included studies, which covers seven domains of potential bias (Figure 2). Dellavia *et al.* [14] showed low risk of bias in all domains except for "Bias in the measurement of outcomes," where the risk was rated as serious, which leads to an overall low risk of bias. Goswami *et al.* [15] had severe risk in "Bias due to confounding," "Bias in the selection of participants," and "Bias in the classification of interventions," but low in all other domains; thus, the overall was at low risk of bias. Gürcan *et al.* [16] were assigned as moderate in "Bias due to confounding" but had low risk in the remaining domains; thus, it has an overall moderate risk of bias. Nadolinski *et al.* [17] demonstrated serious risk in "Bias in the classification of interventions but at low risk in all domains besides this, hence demonstrating overall low risk of bias".

Präger *et al.* [18] presents evidence of moderate risk concerning "Bias due to confounding," "Bias due to missing data," and "Bias in the measurement of outcomes, while at low risk on all other domains such that an overall moderate risk is at stake. Similarly, Seremidi *et al.* [19] was moderate in "Bias due to confounding" and "Bias in the classification of interventions" but low in all other domains and hence overall was moderate. Sushitha *et al.* [20] had a moderate risk in "Bias in the classification of interventions" but all the other domains were low, so the overall was low risk of bias. Terada *et al.* [21] showed moderate risk in "Bias due to confounding," with low risk in all other domains, which finally resulted in an overall moderate risk of bias.

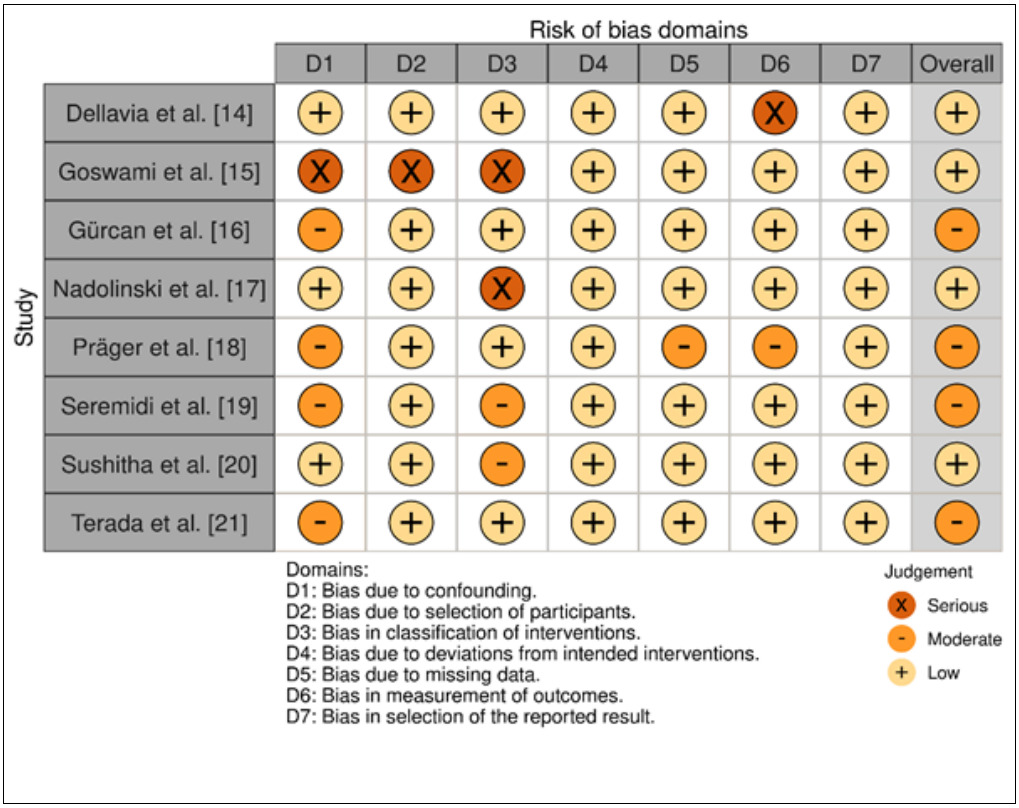

Fig 2: Bias assessment using the ROBINS-I too1

Table 2: Oral findings associated with ED as observed across the included studies

Author ID	Subtype of ED	Genetic Mutation Identified	Detailed Dental Phenotype	Morphological Characteristics	Maxillofacial Parameters	Salivary Gland Function	Periodontal/Mucosal Indicators	Radiographic Findings	Conclusion Assessed
Dellavia *et al*. [14]	Hypohidrotic ED	Mutations in EDA gene	Hypodontia, severe anodontia in some cases	Conical incisors, taurodontic molars	Reduced growth in facial dimensions, narrow palate	Normal salivary gland function	NA	3D imaging and radiographs: reduced bone growth	Prosthetics enhanced growth trajectory
Goswami *et al*. [15]	Hypohidrotic and Hidrotic ED	Not detailed	Multiple missing teeth, uneven distribution	Conical teeth, hypoplastic molars	Decreased alveolar bone height, flat ridges	Reduced salivary flow in some cases	NA	Panoramic radiographs: agenesis of teeth	Improved mastication, speech, aesthetics
Gürcan *et al*. [16]	Various Subtypes of ED	Not detailed	Hypodontia, delayed eruption, missing lateral incisors	Conical teeth, enamel hypoplasia	Vertical facial height reduction, thin alveolar crest	Reduced in some cases, not absent	Good periodontal and mucosal health	Panoramic radiographs	Prosthetic rehabilitation improved aesthetics
Nadolinski *et al*. [17]	ED Subgroup	Mutations in EDA, PAX9	Oligodontia, hypodontia common	Conical and missing molars	Narrow maxillary arch, low bone height	No salivary gland data	NA	Panoramic radiographs: detailed missing teeth	Orthodontics and prosthetics addressed gaps
Präger *et al*. [18]	Hypodontia/Oligodontia	Not detailed	Missing 2-26 teeth, frequent lateral incisor absence	Conical anterior teeth, malformed molars	Hypoplastic alveolar ridge, delayed eruption	NA	NA	Panoramic and CBCT: widespread agenesis	Orthodontic and prosthetic approaches
Seremidi *et al*. [19]	Severe oligodontia	Not detailed	Missing >6 teeth, irregular distribution	Conical primary teeth, taurodontic molars	Reduced vertical dimension, flat ridges	Reduced salivary flow, dry mucosa	NA	Panoramic X-rays: agenesis of teeth	Prosthetic rehabilitation effective
Sushitha *et al*. [20]	Hypohidrotic ED	EDA, EDAR mutations	Missing teeth, no teeth in some cases	Conical in younger sibling, hypoplastic teeth	Frontal bossing, hypoplastic alveolar bone	NA	NA	Panoramic X-rays: missing teeth	Consanguinity-related inheritance suggested
Terada *et al*. [21]	X-linked Hypohidrotic ED	Mutations confirmed in 2 cases	Severe hypodontia/anodontia, small teeth	Conical incisors, abnormal molar shapes	Reduced maxilla size, narrow palate	Significantly low salivary flow	Normal mucosal health	3D scans, lateral cephalograms	Challenges in prosthesis fitting due to anatomy

Discussion

Thematic analysis of the review's findings

Gürcan *et al*. [16] had similar results regarding hypodontia and delayed eruption patterns as Präger *et al*. [18], but the latter study specifically reported malformed molars and hypoplastic alveolar ridges as common features. Salivary gland dysfunction was reported inconsistently; for example, diminished salivary flow was found in Goswami *et al*. [15] and Seremidi *et al*. [19], but normal gland function was reported by Dellavia *et al*. [14]. These variations indicate variation in systemic effect among populations under study.

While all studies highlighted the functional and psychosocial burdens of ED, those with larger sample sizes, such as Nadolinski *et al*. [17], provided broader population-level insights, which contrasted with the case-specific focus of Seremidi *et al*. [19] and Sushitha *et al*. [20]. The latter studies detailed individual presentations of severe phenotypes but lacked generalizability. Prosthetic and orthodontic interventions were more than frequently discussed in research studies, with early rehabilitation providing benefits in promoting craniofacial growth and improving aesthetics as well as function in the Dellavia *et al*. [14] and in the Terada *et al*. [21].

Regarding the differences, the number of subtypes of ED studied in the studies differs, while some, such as Dellavia *et al*. [14], focused on genetic mutations like EDA, others, including Gürcan *et al*. [16] and Nadolinski *et al*. [17], did not specify specific genetic findings. In addition, there was a concentration on advanced imaging modalities including CBCT in Dellavia *et al*. [14] and Terada *et al*. [21]. However, this was discussed less often in smaller-sized studies like Seremidi *et al*. [19]. In summary, studies emphasized the heterogeneity of ED phenotypes and provided a call for more specific diagnostic and therapeutic interventions by individualization.

Comparison of findings with other studies

This review found congruence with those studies put together by Cerezo-Cayuelas *et al*. [22], especially about the necessity for orthodontic and dentofacial orthopedic treatment of patients with ED. These reviews have made light upon early prosthetic and orthodontic treatment in an effort to correct functional as well as aesthetic impairment due to hypodontia, anodontia, and craniofacial anomalies. The frequent use of fixed and removable appliances, palatal expanders, and orthognathic surgery in Cerezo-Cayuelas *et al*. [22] was consistent with our findings, which underscores the importance of individualized rehabilitation strategies for improving mastication, speech, and aesthetics.

The genetic etiology of ED as reported by Wright *et al*. [23] also closely resembled our findings, as both reviews pointed out the central role of mutations in genes such as EDA, EDAR, and PAX9. Moreover, the classification of ED subtypes according to clinical and molecular characteristics is consistent with our observation of varied phenotypic expressions across populations and subtypes.

The consistent dental manifestations across Arcuri *et al*. [25], Garrocho-Rangel *et al*. [24] Zare *et al*. [26], Dev *et al*. [27], and our review include hypodontia, oligodontia, and conical or pegged teeth. Both Zare *et al*. [26] and the present review highlighted the pervasive craniofacial appearance of frontal bossing and reduced alveolar ridge height, along with diminutive vertical facial dimension, in contributing to functional impairments feeding, chewing, and speaking.

The multidisciplinary approach advocated by Garrocho-Rangel *et al*. [24] and Dev *et al*. [27] resonated with the conclusions of our review, which highlighted the necessity of integrating pediatric dentists, orthodontists, prosthodontists, and other specialists for effective management of ED. Both reviews stressed the importance of early intervention and counselling to mitigate psychosocial impacts and improve the quality of life for patients and their families. Although our review focused on the early stimulation of craniofacial growth, Cerezo-Cayuelas *et al*. [22] reported that most of the treatments described in their review were aimed at correcting mispositioning and jaw asymmetries rather than growth promotion. This distinction highlights the need for evidence-based treatment protocols that aim for early functional rehabilitation. Although our study accepted that genetic mutations existed in ED, the proposed detailed classification based on molecular pathways was not a priority for the analysis carried out in our review as suggested by Wright *et al*. [23].

Advanced implant technologies such as zygomatic implants, described by Arcuri *et al*. [25], were not within the focus of our review. Though our findings pointed out the traditional prosthetic and orthodontic interventions, Arcuri *et al*. [25] presented the alternatives to bone grafting with digital implant planning and zygomatic implants for the patients who had severe alveolar deficiencies. Garrocho-Rangel *et al*. [24] had specifically focused on EEC syndrome, while the present review encompasses a wide variety of ED subtypes, like hypohidrotic and hidrotic forms. Garrocho-Rangel *et al*. [24] thereby restricted the generalisability of their findings with this narrow focus.

Prevalence of ED

The prevalence of ED syndromes is different in various studies and populations [28], and estimates indicate it occurs approximately in 6 to 9 per 10,000 individuals [29]. Hypohidrotic ED is the most common subtype of ED and has been reported to affect about 1 in 5,000 to 10,000 live births, though its prevalence is also estimated within the range of 1 to 9 per 100,000 individuals [30].

Genetic Predispositions and Key Characteristics

HED is mainly as a consequence of mutations of genes, which encode for the EDA/NFKappaB signaling pathway such as the EDA, EDAR, EDARADD, WNT10A, involved in development of ectodermal tissue. Of all cases, mutations are genetic in origin and can account for about 90% [31]. HED mainly causes defects in all structures related to the ectoderm, including skin, hair, nails, sweat glands, and teeth. Characteristic phenotypic features are seen in the affected patients, including sparse, thin hair on the scalp and body (hypotrichosis) and pale, smooth skin. The face may present with sunken cheeks, a saddle nose, frontal bossing, prominent supraorbital ridges, hyperpigmentation around the eyes, and everted lips. Lower third of the face can be underdeveloped; lower facial height could be diminished, leading to an aged appearance. In some cases, hyperkeratosis may be present on the palms and soles. Women may also have hypoplastic or absent mammary glands [32-33].

X-Linked Hypohidrotic ED

The most common type of HED, which was previously referred to as Christ-Siemens-Touraine syndrome, is caused by mutations or deletions in the ectodysplasin gene and is inherited in an X-linked pattern. The X-linked form (XHED) is regarded as a rare condition with an incidence of about 1 to

9 per 1,000,000 [34]. Affected individuals with XHED are characterized by hypotrichosis, hypohidrosis, and oligodontia. Most individuals suffering from this condition face respiratory problems resulting from mucosal dryness and malfunctioning of the airways [35].

Other Subtypes of ED

Another subtype is anhidrotic ED, characterized by the absence of sweat and sebaceous glands. On the contrary, hidrotic ED, also referred to as Clouston syndrome, is inherited in an autosomal dominant pattern, where these glands develop normally [36]. These subtypes depict the heterogeneity of ED and its associated clinical features.

Orofacial and Functional Impairments

ED is significantly associated with a wide range of orofacial and functional abnormalities. Nearly all patients present with dental anomalies, including hypodontia, oligodontia, or enamel defects. Difficulties in chewing and swallowing are common and can occur in more than 80% of individuals with ED, whereas dry mouth (xerostomia) occurs in nearly half of the cases. Speech impairments and hoarseness are also problems associated with many ED patients, thereby again pointing to the multidisciplinary impact of the disease on oral and systemic health [37-38].

Limitations

This study was exposed to several limitations. Heterogeneity of study designs and sample sizes contributed to variability in the reported outcomes. Moreover, sparse longitudinal data prevented an understanding of developmental changes over time in a detailed manner. The use of small sample-size studies also led to low generalizability and the fact that there are no standard diagnostic criteria for the subtypes of ED that prevented making direct comparisons. Reporting bias may have affected the inclusion of certain findings, particularly regarding less commonly assessed features like salivary gland function and periodontal health.

Clinical Recommendations and Future Implications

Based on the findings, it is recommended that clinicians adopt a multidisciplinary approach for the early management of ED in pediatric patients. Comprehensive diagnostic protocols, including genetic testing and advanced imaging, should be standardized to facilitate accurate diagnosis and treatment planning. Prosthetic and orthodontic rehabilitation should be initiated early to support craniofacial growth, mastication, and aesthetics. Moreover, education for the patient and caregiver regarding psychosocial impacts of ED is the priority of the clinicians. Longitudinal researches are required further to establish long-term outcome studies for therapeutic interventions to formulate guidelines for evidence-based management of this heterogeneous condition.

Conclusion

This systematic review illustrates the diverse oral and craniofacial manifestation of ED in pediatric patients to highlight its profound effects on dental development and craniofacial structures. The highest frequency findings were hypodontia, oligodontia, and conical teeth but variability in presentation emphasized a need for individualized approach to diagnosis and treatment. Early interdisciplinary intervention is recommended due to its importance in reducing functional, aesthetic, and psychosocial challenges encountered in the management of patients with ED.

Conflict of Interest

Not available

Financial Support

Not available

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