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Disturbances of dental development distinguish patients with oligodontia-ectodermal dysplasia from isolated oligodontia

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Structured Abstract

Objective: To investigate phenotypic differences in dental development between isolated oligodontia and oligodontia-ectodermal dysplasia (ED).

Setting and sample population: A total of 129 patients diagnosed with isolated oligodontia and 22 patients with oligodontia as part of ED were eligible.

Methods: The phenotype of dental development was assessed for the frequency of missing a certain tooth, dental age, development of each tooth present, abnormal size and abnormal shape of teeth. The data were analysed building linear, ordinal and logistic regression models.

Results: Compared to patients with isolated oligodontia, patients with oligodontia-ED missed more frequently central incisors and second molars in both jaws, and lateral incisors in the mandible ($P < .05$). Oligodontia-ED was associated with delayed development of the permanent dentition ($\beta = -0.10$; 95% CI: −0.17, −0.03). Specifically, the maxillary teeth: right central incisor, right lateral incisor, right second premolar and left second premolar were delayed approximately from 2 to 4 developmental stages. In addition, the left mandibular second premolar was 3 developmental stages delayed. Abnormal shape of teeth was 7 times more evident in patients with oligodontia-ED compared to patients with isolated oligodontia (OR = 6.54; 95% CI: 2.34, 18.28). The abnormal size of teeth was not a distinctive characteristic for oligodontia-ED.

Conclusions: Oligodontia-ED distinguishes from isolated oligodontia by more disturbances in dental development. The abnormal shape of incisors and canines in a patient with oligodontia can raise suspicions for accompanying ectodermal abnormalities.

KEYWORDS
ectodermal dysplasia, oligodontia, tooth development

1 | INTRODUCTION

Dental agenesis is the most common anomaly of dental development in humans with a prevalence that varies from 2.6% to 11.3% in different populations.[1,2] According to the number of missing teeth, dental agenesis is classified as hypodontia, oligodontia and anodontia.[3-5] Oligodontia is the developmental absence of 6 or more teeth, excluding the third molars.[6] Oligodontia is observed approximately in 0.14% of the general population,[7] and specifically in the Dutch population, the prevalence of oligodontia is 0.08%.[8] Based on the genetic

evidence, oligodontia is caused by alterations of independent genes or genetic linkages that affect early developmental processes of teeth leading to specific phenotypes.[9] Although oligodontia is usually presented as an isolated trait (OMIM 616724), oligodontia can also be displayed as part of a syndrome.[10] The non-isolated trait of oligodontia is manifested in more than 120 syndromes and quite often it can be the initial sign in diagnosing a patient with a related syndrome.[11]

Ectodermal dysplasia (ED) (OMIM 305100) is the most common group of syndromes associated with oligodontia.[12] ED is characterized by abnormal development of 2 or more ectodermal structures such as hair, skin, nails, salivary and sweat glands, and teeth. Being part of a syndrome, oligodontia presents an extensive phenotype including various dental and craniofacial malformations that require special treatment by an interdisciplinary team of orthodontists, maxillofacial surgeons and prosthodontists.[13]

Genes play an important role in the occurrence of oligodontia and other disturbances of dental development. Particularly, *MSX1*, *PAX9*, *AXIN2*, *EDA*, *EDAR*, *EDARAD,D* and *WNT10A* variants, responsible for isolated oligodontia and oligodontia-ED, are known to be associated also with an aberrant development of the dentition reflected on the structure, number, position and morphology of the teeth.[14-16] Disturbances of dental development that characterize oligodontia refer to the delay of dental development,[17,18] abnormal size (reduced size and short roots of teeth)[19] and abnormal shape (taurodontism, conical shape) of teeth.[20]

However, whether the abnormal features affecting teeth can be distinctive between isolated oligodontia and oligodontia-ED remains an open question as the literature does not share enough insight on the complete phenotype of dental development in the both conditions. Thus, in this study, we aimed to assess the phenotypic differences in dental development between patients with isolated oligodontia and oligodontia as part of ectodermal dysplasia.

2 | MATERIALS AND METHODS

2.1 | Study population

The participants in this study were referred from 1989 to 2016 to 2 medical centres and 1 private orthodontic centre in the Netherlands: Erasmus University Medical Center (Rotterdam), Radboud University Medical Center (Nijmegen) and Orthodontiepraktijk Heerenveen (Heereveen). A total of 182 patients, aged 6-18 years, were identified with oligodontia and had a dental panoramic radiograph (DPR). Thirty-one patients with syndromic oligodontia as part of Down syndrome, clefts or other rare syndromes were excluded from this investigation. The remaining 151 patients (74 females and 77 males) with a median age of 11.30 years (75% range; 8.80-14.18 years) and born from 1975 to 2010 fulfilled the diagnosis selection criteria and were eligible for this study. Patients were classified as manifesting isolated oligodontia (N = 129) and oligodontia as part of ectodermal dysplasia (N = 22). The group of patients manifesting isolated oligodontia was used as the reference group. The utilization of DPRs is in accordance with the general treatment protocol. The study was approved by the Medical Ethics Committee of the Erasmus Medical Center in Rotterdam, the Netherlands (MEC-2017-190). The study conforms to STROBE guidelines.

2.2 | The assessment of oligodontia

Oligodontia was assessed by clinical examination by the dentist or other dental professional and also by detection on the DPRs. A tooth was classified as missing when no sign of formation or calcification was shown on the DPR. Patients with 6 or more congenitally missing teeth, excluding third molars, were diagnosed as oligodontia. Lack of evidence of accompanying abnormal ectodermal features helped the definition of oligodontia as an isolated trait.

2.3 | The assessment of ectodermal dysplasia

During the physical examination, patients identified with 2 or more evident abnormal features of ectoderm (skin, hair, nails and sweat glands) were diagnosed with ectodermal dysplasia and referred to the clinical geneticist. The genetic test confirmed the diagnosis of ectodermal dysplasia. Informed consents were obtained from the patients or parents.

2.4 | The assessment of dental development

Dental development was defined from each available DPR using the Demirjian method.[21] One experienced examiner (B. D) determined 1 of the 8 developmental stages (A, B, C, D, E, F, G and H) for each present tooth in all quadrants. Dental age was calculated for each patient referring to developmental stages of teeth in the left quadrant. As patients missed quite often teeth of the left mandibular quadrant, developmental stages of the missing teeth were ascertained from a combined method.[22] This method consists of assessing the developmental stage of a missing tooth in the lower left quadrant from the corresponding right mandibular tooth or from the corresponding maxillary tooth (when the tooth was missing in both sides of the mandible). In case no corresponding tooth was present, regression equations developed by Nystrom et al were applied.[23] These equations take into account the development of the remaining teeth in the lower left quadrant, age and sex of the patient to calculate dental age. Obtained stages of development were used to calculate the dental maturity score by summing up the weighted scores given to every tooth of the lower left quadrant.[24] Finally, the Dutch dental age standard for boys and girls was used to convert the dental maturity score into dental age.[24] Due to non-normal distribution, dental age was firstly log transformed and further used in the statistical analysis. To obtain a better approach of dental phenotype of the patients, we performed additional measurements. Abnormal shape of teeth including taurodontism, conical and pinned canines, notched incisors and abnormal size of teeth including microdontia, thin and short roots was noted when detected on DPRs and intraoral pictures. We counted dental fillings as a proxy of dental caries and considered as a potential confounder.

2.5 | Statistical analysis

The difference in dental age between isolated oligodontia and oligodontia-ED was investigated using linear regression analysis in 2 consecutive models. Model 1 was adjusted for age. Model 2 was additionally adjusted for the number of missing teeth and number of filled teeth. The difference in the development of each present maxillary and mandibular tooth between isolated oligodontia and oligodontia-ED was analysed using 1 ordinal regression model, adjusted for age, sex, number of missing teeth and number of filled teeth. The same analysis was performed to investigate whether the agenesis of a certain tooth influenced the development of the corresponding tooth in the other jaw. The difference in the abnormal shape of teeth (presence of shape abnormalities or not) between isolated oligodontia and oligodontia-ED was investigated using the binary logistic regression analysis in 2 consecutive steps. Model 1 was adjusted for age and sex. Model 2 was additionally adjusted for the number of missing teeth and number of filled teeth. The same analysis was performed to study the difference in the abnormal size of teeth (presence of size abnormalities or not) between isolated oligodontia and oligodontia-ED. Age, the number of missing teeth and the number of filled teeth were included in the linear regression models based on previous literature or a change of >10% in effect estimates. As sex is taken in consideration when calculating dental age, we used sex as a potential confounder only in the ordinal regression models and binary logistic regression models. All statistical analyses were performed using statistical software Statistical Package for Social Sciences version 21.0 (SPSS Inc., Chicago, IL, USA).

3 | RESULTS

3.1 | General characteristics

The general description of the study population is presented in Table 1. There was no difference in age, sex, abnormal size of teeth and number of filled teeth between the ED patients and isolated oligodontia patients. The number of missing teeth was not statistically significantly different between boys and girls ($P = .771$). The difference between the chronological age and dental age was 1.02 years in patients with isolated oligodontia and 2.88 years in patients with ED. Patients with oligodontia-ED had statistically significantly lower dental age, more missing teeth and were more frequently detected with abnormal shape of teeth than patients with isolated oligodontia.

3.2 | Patterns of oligodontia

The distribution of the missing teeth is presented in Table 2, and the most common patterns of isolated oligodontia and oligodontia-ED are presented in Table 3.

3.2.1 | Isolated oligodontia

The lower second premolars (35, 78.3%; 45, 74.4%), the upper second premolars (15, 72.1%; 25, 69.0%) and the upper lateral incisors (12, 65.9%; 22, 64.3%) were most frequently missing. The upper central incisors (11, 31%; 21, 3.9%), the lower first molars (36, 10.9%; 46, 11.6%) and the upper first molars (16, 14.7%; 26, 12.4%) were less frequently missing.

3.2.2 | Ectodermal dysplasia

The lower central incisors (31, 81.2%; 41, 81.8%), second premolars (15, 68.2%; 25, 68.2%; 35, 68.2%; 45, 72.7%), second molars (17, 72.7%; 27, 68.2%; 37, 68.2%; 47, 68.2%) and lateral incisors (12, 63.6%; 22, 72.7%; 32, 63.6%; 42, 68.2%) were most frequently missing. The upper first molars (16, 13.6%; 26, 13.6%) and lower canines (33, 18.2%; 43, 13.6%) were less frequently missing. The frequency of missing the central incisors ($P < .01$), the lower lateral incisors

TABLE 1 General characteristics of the study population (N = 151)

	Isolated oligodontia (N = 129)	Oligodontia-ED (N = 22)	*P*-value
Age	11.32 (8.81-14.05)	10.98 (7.19-14.51)	.918
Sex (N; %)			
Females	65 (50.0)	9 (41.0)	.278
Males	64 (50.0)	13 (59.0)	
Number of missing teeth	10 (6-17)	14 (6-22)	*<.001*
Dental age	10.30 (7.55-12.48)	8.10 (5.40-11.56)	*.012*
Abnormal size of teeth (N; %)	24 (18.6)	7 (31.8)	.130
Abnormal shape of teeth (N; %)	22 (17.1)	14 (63.6)	*<.001*
Number of filled teeth	0 (0-2)	0 (0-0)	.876

N, number of participants; values are percentages for dichotomous variables or medians (75% range) for ordinal and continuous variables with a skewed distribution. Differences were tested using the Mann-Whitney *U* non-parametric test for continuous variables and Chi-squared test for categorical variables; $P < .05$ is considered statistically significant and presented in italic font.

TABLE 2 Developmental stages and distribution of agenetic teeth

Developmental stages (0-8) of teeth				Distribution of agenetic teeth		
FDI Code	Isolated oligodontia (N = 129)	Oligodontia-ED (N = 22)	*P*-value	Isolated oligodontia (N = 129)	Oligodontia-ED (N = 22)	*P*-value
11	8 (7-8)	8 (0-8)	*.001*	4 (3.1)	5 (22.7)	*.004*
12	0 (0-8)	0 (0-8)	.632	85 (65.9)	14 (63.6)	.507
13	6 (0-8)	6 (0-8)	.912	36 (27.9)	5 (22.7)	.414
14	0 (0-7)	0 (0-6.3)	.557	71 (55.0)	13 (59.1)	.454
15	0 (0-6)	0 (0-5.1)	.913	93 (72.1)	15 (68.2)	.442
16	8 (0-8)	8 (0-8)	.169	19 (14.7)	3 (13.6)	.597
17	5 (0-7)	0 (0-7.1)	*.011*	43 (33.3)	16 (72.7)	*.001*
21	8 (7-8)	8 (0-8)	*.028*	5 (3.9)	5 (22.7)	*.006*
22	0 (0-8)	0 (0-8)	.460	83 (64.3)	16 (72.7)	.306
23	6 (0-8)	6 (0-8)	.577	31 (24.0)	6 (27.3)	.464
24	0 (0-8)	1.5 (0-7.1)	.817	71 (55.0)	11 (50.0)	.416
25	0 (0-6)	0 (0-5.1)	.829	89 (69.0)	15 (68.2)	.560
26	8 (0-8)	8 (0-8)	.307	16 (12.4)	3 (13.6)	.549
27	5 (0-7)	0 (0-7)	*.005*	46 (35.7)	15 (68.2)	*.004*
31	7 (0-8)	0 (0-8)	*.004*	62 (48.1)	18 (81.2)	*.003*
32	8 (0-8)	0 (0-8)	*.003*	39 (30.2)	14 (63.6)	*.003*
33	7 (0-8)	7 (0-8)	.805	20 (15.5)	4 (18.2)	.478
34	5 (0-8)	0 (0-7.1)	.161	49 (38.0)	12 (54.5)	.110
35	0 (0-5)	0 (0-5.1)	.471	101 (78.3)	15 (68.2)	.218
36	8 (7-8)	7.5 (0-8)	*.012*	14 (10.9)	5 (22.7)	.117
37	4 (0-7)	0 (0-7)	*.023*	55 (42.6)	15 (68.2)	*.023*
41	0 (0-8)	0 (0-8)	*.009*	66 (51.2)	18 (81.8)	*.006*
42	8 (0-8)	0 (0-8)	*.008*	48 (37.2)	15 (68.2)	*.007*
43	7 (0-8)	7 (0-8)	.650	21 (16.3)	3 (13.6)	.522
44	5 (0-8)	0 (0-8)	.131	47 (36.4)	12 (54.5)	.086
45	0 (0-6)	0 (0-5.1)	.937	96 (74.4)	16 (72.7)	.526
46	8 (6.3-8)	8 (0-8)	.208	15 (11.6)	4 (18.2)	.290
	4 (0-7)	0 (0-7)	*.017*	53 (41.1)	15 (68.2)	*.017*

FDI, World Dental Federation 2-digit tooth notation; differences in developmental stages of each tooth between oligodontia-ED and isolated oligodontia are tested by Mann-Whitney *U* non-parametric test; differences in agenesis of each tooth between oligodontia-ED and isolated oligodontia are tested by Chi-squared test, and significant *P*-values are presented in italic font.

($P < .01$) and the second molars ($P < .05$) was statistically significantly higher in ED patients compared to isolated oligodontia patients.

3.3 | Differences in dental development between isolated oligodontia and oligodontia-ED

3.3.1 | Dental age

As part of ED, oligodontia was associated with a delayed development of the permanent dentition in Model 1 (β = −0.17; 95% CI: −0.25, −0.09). The effect estimate decreased in Model 2 (β = −0.10, 95% CI: −0.17, −0.03), however, the association remained statistically significant (Table 4).

3.3.2 | The development of each present tooth

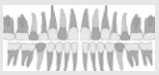
Maxillary teeth

As shown in Figure 1, the ordinal regression analysis revealed a statistically significant association of oligodontia as part of ED with the delayed developmental stages of the right central incisor (β = −1.65; 95% CI: −3.03, −0.27), the right lateral incisor (β = −3.53; 95% CI: −6.34, −0.73), the right second premolar (β = −3.19; 95% CI: −5.11, −1.28) and the left second premolar (β = −2.32; 95% CI: −4.07, −0.57).

Mandibular teeth

The ordinal regression analysis (Figure 1) showed a statistically significant association of oligodontia as part of ED with the developmental stages of the left second premolar (β = −2.93; 95% CI: −4.93, −0.93).

TABLE 3 The frequency of oligodontia patterns

	TAC	Missing teeth (FDI)	Illustration	N (%)
Isolated oligodontia				
Maxilla	48	15, 14, 24, 25		12 (9.3)
	52	15, 14, 12, 22, 24, 25		13 (10.1)
	The others			104 (80.6)
Mandible	32	45, 35		14 (10.9)
	48	45, 44, 34, 35		13 (10.1)
	The others			102 (79.0)
Overall dentition	68	15, 12, 22, 25, 45, 35		5 (3.9)
	96	15, 14, 24, 25 45, 44, 34, 35		5 (3.9)
	The others			119 (92.2)
Oligodontia-ED				
Maxilla	52	15, 14, 12, 22, 24, 25		2 (9.1)
	180	17, 15, 14, 12, 22, 24, 25, 27		2 (9.1)
	The others			18 (81.8)
Mandible	6	42, 41, 31, 32		3 (13.6)
	182	47, 45, 44, 42, 41, 31, 32, 34, 35, 37		2 (9.1)
	The others			17 (77.3)

FDI, World Dental Federation 2-digit tooth notation, TAC—tooth agenesis code, ED—ectodermal dysplasia, crown of the missing teeth are illustrated in dark grey colour[27]; patterns that were less frequent are presented as "the others," patterns that were present only in 1 patient are not presented.

Antagonists of agenetic teeth

The results of the studied association between agenesis of a certain tooth and development of the correspondent in the other jaw are shown in Figure 2.

3.3.3 | The abnormal shape of teeth

As shown in Table 5, oligodontia-ED was associated with the abnormal shape of teeth (OR = 8.51; 95% CI: 3.14, 23.03) in Model 1. The association of non-isolated oligodontia with the abnormal shape of teeth remained still statistically significant ($P < .001$) in Model 2, however, the effect estimate decreased (OR = 6.54; 95% CI: 2.34, 18.28).

3.3.4 | The abnormal size of teeth

The effect estimates obtained in Model 1 and Model 2 of the logistic regression analysis did not present distinctive differences between isolated oligodontia and oligodontia-ED (Table 5). Considering all the possible confounders in Model 2, oligodontia as part of ED (OR = 2.16; 95% CI: 0.67, 7.00) was not statistically significantly associated with the abnormal size of teeth.

4 | DISCUSSION

In this study, we investigated the phenotypic differences in dental development between patients with isolated oligodontia and oligodontia as part of ectodermal dysplasia. Patients with oligodontia and ectodermal dysplasia showed disturbances in dental development the most. The disturbed development of teeth was mainly expressed in the higher frequency of missing the central incisors and second molars in both jaws, and the lower lateral incisors. Furthermore, a delayed maturation of the permanent dentition of approximately 10 months-1 year and a half was shown when compared with isolated oligodontia patients. Specifically, the development of the maxillary teeth, such as right central incisor, right lateral incisor, right second premolar and left second premolar, was around 2-4 stages delayed. As regarding to the mandibular teeth, the left second was approximately 3 stages delayed

TABLE 4 The association between oligodontia-ED and dental age

	Model 1			Model 2		
	β	95% CI	*P*-value	β	95% CI	*P*-value
Ectodermal dysplasia (isolated oligodontia; ref.)	−0.17	−0.25, −0.09	<.001	−0.10	−0.17, −0.03	*.008*

β, regression coefficients, CI, confidence interval, ref., reference; significant *P*-values are presented in italic font.
Dental age was calculated if both matching mandibular teeth were missing by scoring them: a—as a developmental stage of the (left) matching maxillary tooth, b—as a developmental stage calculated from regression equations developed by Nystrom et al. (2000).[23]
Model 1: adjusted for age.
Model 2: was additionally adjusted for number of missing teeth and number of filled teeth.

in development when being present. Abnormal shape of teeth was approximately 7 times more evident in patients with oligodontia-ED than in patients with isolated oligodontia.

Our findings were consistent with the literature, as patients with oligodontia-ED are expected to show more disturbances in dental development than patients with isolated oligodontia due to the higher occurrence of dental anomalies affecting the number, size and shape of teeth.[25,26] Oligodontia as part of a syndrome is characterized by more agenetic teeth than isolated oligodontia,[26,27] shown in our study as well. The lower second premolars and upper lateral incisors are recognized as the most frequent congenitally missing teeth.[2,28] Consistently, second premolars and lateral incisors were among the most prevalent missing teeth in both groups of isolated oligodontia and oligodontia-ED. Beside the common agenetic teeth, the frequency of missing the central incisor and second molar was distinctive for ED patients. While the agenesis of the central incisor indicates oligodontia of 9 or more teeth, the agenesis of the second molar is to our best knowledge not previously mentioned to distinguish patients with oligodontia-ED[15]; raising the question whether the agenetic second molar could be potentially a phenotypic indicator of ED. The teeth noted as the most prevalent missing showed also delayed developmental stages when present. However, the delay in maturation of all permanent teeth was a general trend in patients with oligodontia-ED. We obtained more significant differences in the development

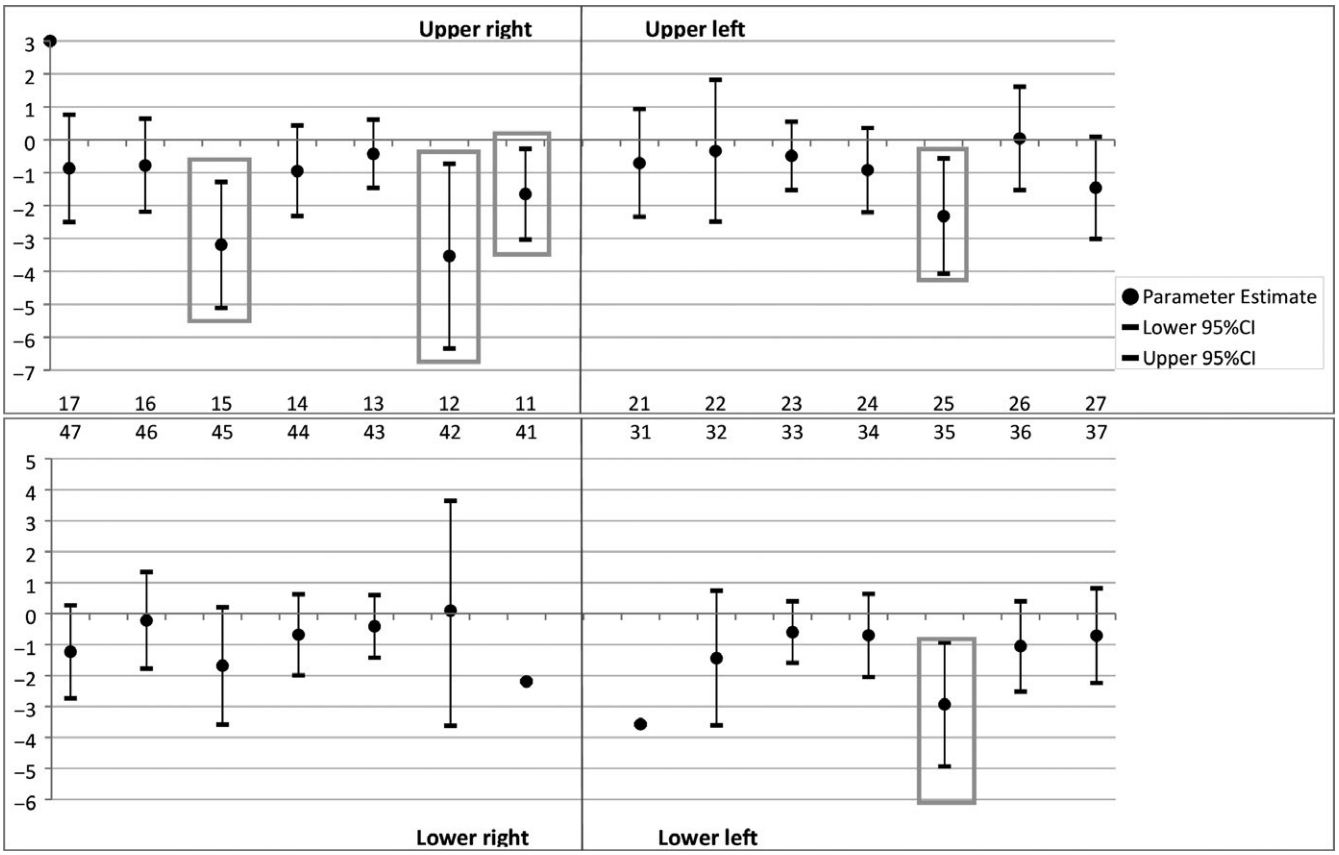


FIGURE 1 The associations of oligodontia-ED with developmental stages for each tooth present. The ordinal regression model was fully adjusted for age, sex, number of missing teeth and number of filled teeth; the statistically significant associations are presented in the grey squares

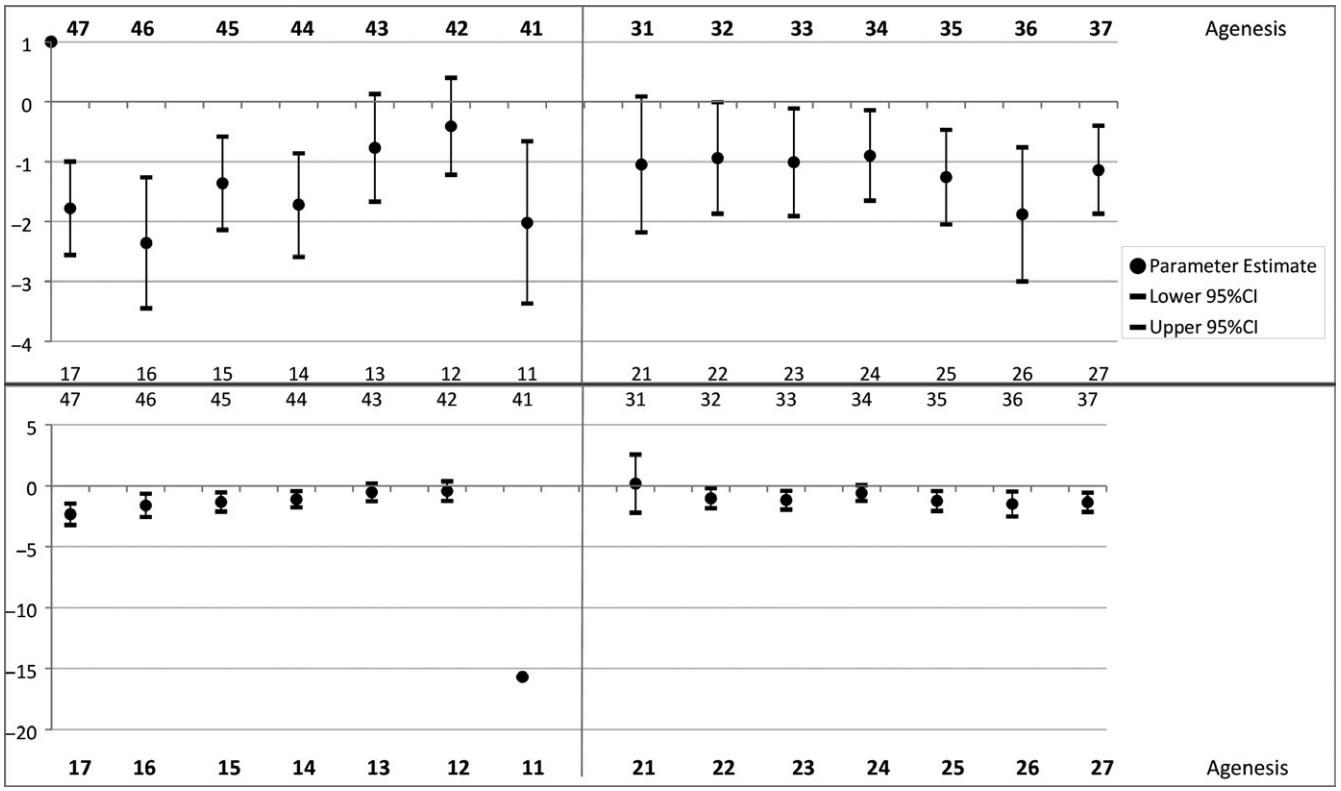


FIGURE 2 The association of the agenesis of a certain tooth with the development of its antagonist. The ordinal regression model was fully adjusted for age, sex, number of missing teeth and number of filled teeth; the statistically significant estimates do not cross the reference axis (zero)

TABLE 5 The associations of oligodontia-ED with abnormal size and abnormal shape of teeth

	Model 1			Model 2		
	OR	95% CI	*P*-value	OR	95% CI	*P*-value
Abnormal shape (isolated oligodontia; ref.)	8.51	3.14, 23.03	*<.001*	6.54	2.34, 18.28	*<.001*
Abnormal size (isolated oligodontia; ref.)	1.19	0.79, 6.20	.132	2.22	0.73, 6.75	.160

OR, odds ratios, CI, confidence interval, ref., reference; significant *P*-values are presented in italic font.
Model 1: adjusted for age and sex.
Model 2: was additionally adjusted for number of missing teeth and number of filled teeth.

of maxillary teeth than in the development of mandibular teeth. Considering the trend of mandibular teeth being more frequently agenetic than maxillary teeth, a distinguished delay of development in mandibular teeth was expected.[28] A significant association between agenesis of maxillary second premolar and the corresponding mandibular second premolar (antagonist) has been previously shown.[29] When present, the antagonists of the most common missing teeth in patients with oligodontia tended to present lower developmental stages, linking the agenesis of a certain tooth with the delayed development of its antagonist. As expected, patients with ED had a significant higher frequency of malformed teeth mainly expressed for maxillary canines and central incisors. The conical shape of the crown in canines and notched marginal edge of incisors were notable in 64% of oligodontia-ED patients. The shape of dental crown is determined by the shape of the enamel layer deposited upon the dentin layer.[30,31] As the only dental tissue originating from ectoderm, enamel is the main bridge that links disturbed dental development with ectodermal dysplasia. Abnormal formation and mineralization of enamel can influence the shape of dental crown and the developmental stages of the affected teeth due to calcification process.[32] Thus, more malformed teeth and more delayed developmental stages can distinguish patients with oligodontia as part of ED from patients with isolated oligodontia. Smaller tooth size characterizes patients with isolated oligodontia and also patients with oligodontia-ED.[19,28,33] In our study, the abnormal size of teeth was not a distinctive characteristic of oligodontia-ED compared to isolated oligodontia.

Clinical reports describe isolated oligodontia as a condition that can be associated with appearance of abnormal ectodermal features

from hair, nails and sweat glands.[15] Hence, the distinction of isolated oligodontia from non-isolated oligodontia becomes a common clinical concern in patients with ectodermal dysplasia. Recently, genetic mutations of *WNT10A* implicated in with the condition of isolated oligodontia are shown to be associated also with ED.[15] Thus, a proper differentiation between the both conditions is a necessity. The investigation of genotype-phenotype associations in patients with isolated oligodontia and oligodontia-ED would be the best solution to achieve the distinction of 1 condition from the other in a clinical and literary perspective. The lack of genetic analysis in isolated oligodontia limited us to attach information on genetic traits to each patient. Furthermore, we could not obtain additional information about possible presence of abnormal ectodermal symptoms affecting salivary secretion, hair, skin and nails in isolated oligodontia. However, to help the distinction of isolated oligodontia from oligodontia-ED especially when a genetic test is not performed and the abnormal features of ectoderm are not evident in the clinical examination, we assessed the dental development phenotype for each patient and additionally defined the specific dental differences between the both conditions.

In the current study, the measurements on dental development are based on DPRs as the most reliable instrument to assess the mineralization stages of teeth. A DPR is an important diagnostic tool in the dental clinical practice; however, detailed information on abnormal size and shape of teeth cannot be precisely estimated only form DPRs. In addition, we used the intraoral pictures to extract the most accurate information.

Although oligodontia is a rare congenital anomaly, it carries on an esthetical, functional, psychological and financial burden for all the patients.[34,35] This study includes only 22 cases of ectodermal dysplasia known as the most common group of syndromes where oligodontia is manifested as a non-isolated trait, leaving in shadow many other rare syndromes. However, syndromic oligodontia is reported as a rare condition, decreasing the power of our study due to the small sample size and also limiting the performance of other studies in this specific group of patients.

Our findings suggest that oligodontia-ED distinguish from isolated oligodontia by more agenetic second molars, evident abnormal shape of incisors and canines, and 1 year delayed development of the present teeth, reflected in lover developmental stages of the maxillary premolars the most. In conclusion, phenotypic differences in dental development exist between isolated oligodontia and oligodontia-ED and should be recognized to facilitate the differential diagnosis between the both conditions.

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CONFLICT OF INTEREST

The authors declare that they have no conflict of interest.

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