

Dental development and molar root length in children with cleidocranial dysplasia

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Abstract

Cleidocranial dysplasia (CD) is an autosomal dominant skeletal disorder with characteristic dental findings of numerous supernumerary teeth and noneruption of permanent teeth. This investigation compared the dental development and root lengths of the mandibular first permanent molar in 11 CD patients with those of 22 healthy, normal children matched for race, age and sex. The results showed that children with CD experienced a delay in dental development of approximately 3 years compared with normal children ($P < 0.05$). In addition, the root lengths of the mandibular first permanent molar were significantly longer than those of the comparison children (17.8 ± 1.6 mm vs 13.6 ± 1.2 mm ($P < 0.001$). This study thus revealed two significant clinical features of CD: 1) severe delay in dental development, and 2) excessive root lengths of mandibular permanent first molars. These features may be important in the pathogenesis of delayed dental eruption observed in this disorder. (Pediatr Dent 17:101-5, 1995)

Cleidocranial dysplasia (CD) is a well-known syndrome with characteristic abnormalities of the skeleton and teeth.^{1,2} An autosomal dominant mode of transmission has been most commonly reported, although sporadic cases and autosomal recessive cases also have been documented.³

Typical features of CD include absence or reduced size of one or both clavicles, brachycephaly with parietal and frontal bossing, hypoplastic maxilla and zygoma, and relative mandibular prognathism.⁴⁻⁶ Other skeletal abnormalities include delayed closure of the fontanels, pubic symphysis and coxa vara, as well as anatomical changes in the vertebrae and phalanges.^{2,3,7}

Oral manifestations often reported are supernumerary teeth, which may develop continuously over a period of time^{8,9} and noneruption of the permanent teeth.^{5,7,9} In addition, various dental crown and root abnormalities have been documented.¹⁰⁻¹³

To date studies on CD have been mainly case reports with one exception.¹⁴ Of note is the paucity of information on dental growth and development as well as on the interaction of dysplastic bone and the dental tissues. The aim of this investigation was to compare the dental maturity and root development of a group of patients with CD with age- and sex-matched healthy controls of similar race.

Patients and methods

Cleidocranial dysplasia patients

Eleven Caucasian patients (7 females and 4 males) with a definitive medical diagnosis of CD had panoramic radiographs available for study. Five were undergoing dental treatment at the Children's Hospital, Boston, Massachusetts, and another three were referred to one of the authors for treatment at the University of Queensland Dental School, Brisbane, Australia. Two more patient records were provided to the authors from the University of Minnesota (R Kuba) and another from the Children's Dental Hospital, Brisbane, Australia (R Auer).

Comparison patients

For every CD patient, two healthy, normal comparison patients matched for race, sex, and age were selected randomly from the patient records kept at the Department of Dentistry, Children's Hospital, Boston. These comparison children had panoramic radiographs exposed at chronologic ages within 6-12 months of those of CD patients.

Dental age assessment

Panoramic radiographs were used to assess patients' dental ages by employing the method of Demirjian et al.¹⁵ In brief, the individual radiological appearances of the seven permanent teeth on the left side of the mandible were evaluated according to developmental criteria. Each tooth was categorized into one of eight stages. A numerical score for each tooth was obtained from standard references for each developmental stage, and the summed scores on all seven teeth gave a dental maturity score. The dental age of each patient was obtained by comparing the dental maturity score to normal standards. To assess dental maturity, only those patients younger than 16 years of age with panoramic radiographs were included, since dental maturity cannot be accurately assessed once the second permanent molars are completely developed.¹⁵

The dental age assessment was done by one author (WKS). Intraexaminer reliability was established using the radiographs of three normal, healthy children aged 9-12 years, who were not part of the study. Triplicate scoring of the radiographs revealed no statistical differences in the mean scores ($P > 0.1$).

The ready identification of CD cases in the radiographs precluded "blinding" of the examiner while assessing dental ages.

Assessing tooth crown-body and root lengths

The crown-body (CB) and root (R) lengths of the mandibular first permanent molar were obtained from panoramic radiographs of the patients using the method of Seow and Lai.¹⁶ Briefly, the CB of each tooth was determined by measuring the length along the vertical axis of the tooth from a perpendicular line drawn through the occlusal pit to a perpendicular line drawn through the furcation (Fig 1). The R-value was determined along the same axis from the furcation to the root apices. Measurements were taken to the nearest

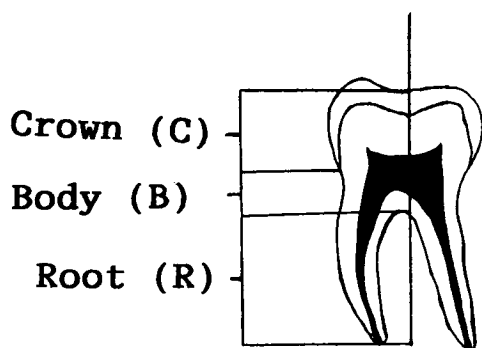


Fig 1. Measurement of the Crown(C)-Body(B) and Root(R) lengths of the mandibular first permanent molar.

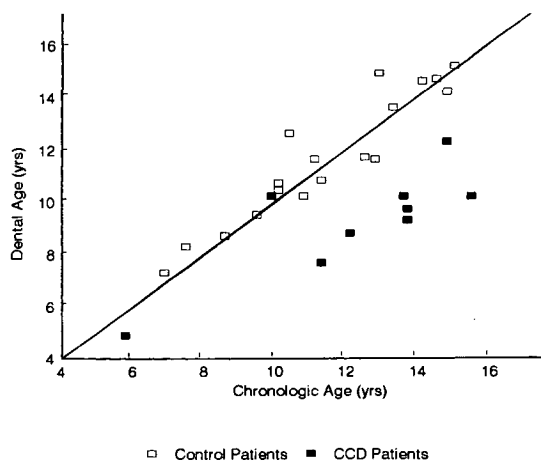


Fig 2. Scattergraph showing the distribution of chronologic and dental ages of CCD and comparison patients. With the exception of one patient, the dental ages of CCD patients were located to the right of the isochrone line (which joins points of identical chronologic and dental ages), indicating delayed dental ages. In contrast, the dental ages of the control patients were scattered close to the isochrone line.

0.1 mm using a digital caliper (Mitutoyo®, Tokyo, Japan). Each tooth was measured three times, and a mean obtained for each measurement. Both mandibular first permanent molars were measured, and means of their measurements were used to analyze the data.

The mandibular first permanent molar was selected for measurement because previous studies¹⁶ showed that there is little distortion of the panoramic radiographic image of this tooth compared with its long-cone radiographic image.

Statistical analysis

The Student's *t*-test was used for statistical analysis of the data.

TABLE 1. COMPARISON OF CHRONOLOGIC AND DENTAL AGES IN CD AND NORMAL PATIENTS

	Chronologic Age (CA) Mean ± SD (yrs)	Dental Age (DA) Mean ± SD (yrs)	Difference (DA-CA) Mean ± SD (yrs)	P-value
Cleidocranial dysplasia				
(N = 9)	12.3 ± 2.8	9.3 ± 1.9	3.0 ± 1.3	<0.05
Range	5.8 - 15.5	4.9 - 12.3	0.3 - 4.4	
Normal				
(N = 18)	11.5 ± 2.4	11.7 ± 2.4	0.2 ± 0.5	>0.1
Range	6.9 - 14.8	7.3 - 14.7	0.8 - 2.0	

Dental ages of the children were assessed using the method of Demirjian et al.¹⁵

The Student's *t*-tests were used for statistical analysis of the data.

TABLE 2. CROWN-BODY (CB) AND ROOT (R) LENGTHS OF PATIENTS WITH CLEIDOCRANIAL DYSPLASIA COMPARED WITH AGE- AND SEX-MATCHED NORMAL CHILDREN

	Cleidocranial Dysplasia (N = 11)	Normal (N = 22)	Difference	P-value
Crown-body (CB) length				
Mean mm ± SD	11.4 ± 1.3	12.2 ± 0.7	0.8 ± 1.3	>0.1
Range (mm)	10.5 - 14.0	10.5 - 13.0		
Root (R) Length				
Mean mm ± SD	17.8 ± 1.6	13.6 ± 1.2	4.2 ± 1.6	<0.001
Range (mm)	12.5 - 22.0	9.0 - 15.3		
CB:R ratio				
Mean ± SD	0.6 ± 0.08	0.9 ± 0.03	0.3 ± 0.08	<0.001

The CB and R lengths were assessed from panoramic radiographs using the method of Seow and Lai.¹⁶

The Student's *t*-test was used for statistical analysis of data.

Results

Chronologic and dental ages of children with cleidocranial dysplasia

Nine CD patients had panoramic radiographs taken at ages suitable for dental age assessment. Two other patients aged >16 years at the time of radiographic exposure were excluded from this part of the study. As shown in Table 1, their mean chronologic age at the time of panoramic radiographic examination was 12.3 ± 2.8 years (range 5.8–15.5 years). In contrast, their mean dental age was only 9.3 ± 1.9 years (range 4.9–12.3 years), indicating a severe delay in dental development. The difference between the mean chronologic and dental ages was 3.0 ± 1.3 years (range 0.3–4.4 years) which was statistically significant ($P < 0.05$).

The mean chronologic age of the normal comparison patients was 11.5 ± 2.4 years (range 6.9–14.8 years), which was not significantly different from that of the CD patients, ($P > 0.1$). Also, their mean dental age was 11.7 ± 2.4 years (range 7.3–14.7), which was not significantly different from their mean chronologic age (11.5 ± 2.4).

Fig 2 shows a scattergraph of dental ages plotted against chronologic ages for CD and comparison patients. The figure clearly shows dental ages of CD patients to the right of the isochrone line, (dental ages were at lower values compared with the corresponding chronologic ages).

Crown-body (CB) and root (R) lengths of the mandibular first permanent molar

Eleven patients with CD had panoramic radiographs suitable for the assessing CB and R.¹⁷ Twenty-two healthy, normal children, matched for race, age, and sex were selected at random for comparison.

As shown in Table 2, the mean CB of the mandibular first permanent molar of patients with CD was 11.4 ± 1.3 mm as compared with 12.2 ± 0.7 mm for the normal children, ($P > 0.1$). This comparison indicated no significant difference in the mean CB values between the two groups.

In contrast, the mean R of the mandibular first permanent molar of patients with CD was 17.8 ± 1.6 mm (range 12.5–22.0 mm) compared with 13.6 ± 1.2 mm (range 9.0–15.5mm) for the normal children (Table 2). The difference in mean R was statistically significant ($P < 0.001$). Also, the CB:R ratio was 0.6 in the CD patients compared with 0.9 in the normal children, ($P < 0.001$).

Correlation of root lengths with developmental delay

For each patient, the mean R of the mandibular first permanent molar was correlated with the corresponding developmental delay observed (Table 3). No association of increased root length with developmental delay was noted ($P > 0.1$).

Discussion

CD is a rare skeletal disorder with significant dental implications. Well-known dental signs of the condition are numerous supernumerary teeth and noneruption of the permanent teeth. In our controlled study, we have demonstrated two additional dental anomalies not widely recognized in this disorder: a marked delay in dental development and excessive root length formation. Although our study of 11 patients constitutes one of the larger series of this rare disorder in the literature, caution must still be exercised in interpreting the results of the relatively small number of patients.

In our investigation, patients with CD showed a mean delay in dental maturity of approximately 3 years compared with race-, age-, and sex-matched healthy, normal comparison children. Our results thus extend those of a previous study by Jensen and Kreiborg,¹⁴ which demonstrated delayed development of permanent teeth and normal development of primary teeth. However, this previous study, although longitudinal in nature, did not include healthy children for comparison, and hence, their results were difficult to interpret. In contrast, in our controlled study, comparison data from normal children provide convincing evidence that CD patients are delayed in their dental development. The normal comparison children in our study, although taken from an institution, showed close correlation of their dental ages with respect to their chronologic ages, indicating that they have growth similar to the normal values established by Demirjian et al.¹⁵

The abnormality in dental development is likely to be related to general skeletal delay in CD. Although this aspect of development has not

TABLE 3. DENTAL MATURITY AND ROOT LENGTHS OF CHILDREN WITH CLEIDOCRANIAL DYSPLASIA

Patient (sex)	Chronologic Age (CA)* (yrs)	Dental Age (DA) (yrs)	Difference (DA-CA) (yrs)	Mean Root Length (\bar{x} 's)(mm)
1 (M)	12.1	8.8	-3.3	19.5
2 (F)	9.9	10.2	0.3	16.5
3 (F)	5.8	4.9	-0.9	n.d.
4 (M)	11.3	7.7	-3.6	12.8
5 (F)	14.8	12.3	-2.5	16.5
6 (F)	15.5	10.2	-5.3	16.7
7 (F)	13.6	10.2	-3.4	17.0
8 (F)	13.7	9.3	-4.4	21.0
9 (F)	13.7	9.7	-4.0	17.5
Mean \pm SD	12.3 ± 2.8	9.2 ± 1.9	-3.1 ± 1.3	17.1 ± 2.2

* Chronologic age indicates age when panoramic radiographic was exposed.

Nine of 11 CD children were included in this part of the study. Two children were excluded in the assessment of dental ages because their panoramic radiographs were exposed after 16 years of age.

Dental ages were assessed using the method of Demirjian et al.¹⁵



Fig 3. Typical panoramic radiographic appearance of patient with cleidocranial dysplasia, showing numerous supernumerary teeth and noneruption of the permanent teeth. Note excessive lengths of the roots of the mandibular first permanent molars.

been researched in CD, affected patients show various abnormalities suggestive of growth changes, such as a prolonged delay in the closure of the anterior fontanel, sagittal, and metopic sutures, and the pubic symphysis.³ In addition, diminished turnover of bone also has been reported in the cranium of CD patients.¹⁸ Thus, the pathogenesis of delay in dental maturity is likely to be a direct manifestation of the general spectrum of skeletal aberrations observed in this condition.

The severe delay in dental development in CD may contribute significantly to the retardation of tooth eruption — a common sign of the condition. Our findings have thus provided another plausible explanation for the pathogenesis of delayed dental eruption in this syndrome. Previous authors have speculated that delayed dental eruption may be the result of physical impediment from numerous supernumerary teeth,^{19,21} or be related to malformation of roots and absence of cellular cementum.^{12, 20-23} Abnormal turnover of bone also has been suggested as a possible etiological factor.¹³ However, while all these previously suggested factors may contribute to retarded eruption, we speculate that the dental development delay is the major contributing factor in retardation of dental eruption.

While a few previous case reports have noted abnormalities in root development in CD such as dilacerations,^{19,21} there is minimal information regarding root length in CD, and objective evaluations of root lengths in affected patients have not been attempted

before. Shafer et al,²⁴ in their textbook on oral pathology stated that the dental roots in CD are “somewhat short and thinner than usual, and may be deformed”. In contrast to their statement, our study has shown that the mean root length of the mandibular first permanent molar is approximately 4 mm greater in patients with CD (Fig 3) compared with normal children. Although previous authors have attributed root anomalies to prolonged mechanical retention in the bone,¹⁹ it was noted in our series of patients that most of the increase in root length occurred after eruption, so that this is not a response to mechanical retention.

Furthermore, we did not find any association of increased root length of the mandibular first permanent molar with increasing severity of developmental dental delay in either the CD or normal patients. Thus, it is likely that the excessive root development in CD results from an intrinsic dental root sheath abnormality associated with the general skeletal dysplasia, rather than from mechanical obstruction. Alternatively, as dental development is also known to be influenced by surrounding bone, it is also possible that excessive root development may have resulted from abnormal interaction with surrounding dysplastic bone.

The presence of numerous supernumerary teeth¹⁴ also supports the theory that abnormal dental development is part of the skeletal aberrations in CD. This anomaly, which is likely to have resulted from incomplete or delayed resorption of the dental lamina,¹⁴ together with our findings further demonstrate the broad spectrum of dental tissue involvement in this unique skeletal dysplasia.

Conclusions

Patients with CD show a dental developmental delay of approximately 3 years compared with normal children, which may be one of the reasons for the retarded eruption of the permanent teeth. In addition, the root lengths of the mandibular first permanent molar of CD children averaged approximately 4 mm longer than molars of healthy children.

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From the Archives

California lifestyle and its effects, circa 1897

An extraordinary prevalence of diseases of the heart and arteries is reported in California, owing, it is thought by the physicians in that locality, to the habit of using such large quantities of intoxicating liquors.

Lancet, 1897

Dealing with an 1879 dental operatory annoyance

The attraction which blood has for flies causes them to gather about the cuspador in a manner that is exceedingly disagreeable both to patient and operator. A little oil of peppermint dropped into the cuspador will lessen its attractions very much. A few drops of coal-oil will equally disgust the pests, and cause them to seek food in other directions.

Dental Cosmos, 1879