

ABSTRACT

Cleidocranial dysplasia – treatment of consequential disturbances to tooth eruption

Background – The purpose of the present study was to report the outcome of dental treatment in a boy with cleidocranial dysplasia (CCD) managed according to the interceptive treatment strategies previously suggested by Jensen and Kreiborg.

Case study – The boy was followed from infancy to adulthood with clinical examinations, photos, orthopantomographic radiographs and roentgencephalometry to monitor craniofacial and dental development and treatment outcome.

The primary dentition was normal. The permanent dentition revealed anomalies in tooth formation, maturation and eruption. He developed 5 supernumerary teeth and severe disturbances of tooth eruption of nearly all teeth. The general treatment principle was to remove supernumerary teeth, primary teeth and bone covering the normal permanent teeth at the time when these teeth had developed about ½ of their final root length to promote spontaneous eruption. Nearly all teeth erupted spontaneously and the need for corrective orthodontic treatment was limited to a relatively short period.

Conclusion – The findings support the suggestion that in CCD an interceptive treatment approach with removal of the obstacles for eruption (supernumerary teeth, primary teeth, and alveolar bone) at the time when the normal teeth have developed about ½ of their final root length can lead to spontaneous eruption of the normal teeth.

Cleidocranial dysplasia: Interceptive treatment of disturbances of tooth eruption

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Cleidocranial dysplasia (CCD) is a rare skeletal disorder with delayed osteoblast differentiation and autosomal dominant inheritance caused by mutations in the *RUNX2* gene (1,2). The main clinical features include short stature, hypoplastic or aplastic clavicles, delayed ossification of calvaria, multiple Wormian bones, wide forehead, midface hypoplasia and dental anomalies; including supernumerary teeth and severely delayed or arrested tooth eruption (1,3). Kreiborg et al. (4) suggested, based on radiographic findings of altered surface remodelling of the jaws that osteoclast differentiation and function are also affected, and Yoda et al. (5) and Lossdörfer et al. (6) found that impaired recruitment of osteoclasts is one of the mechanisms of delay or lack of tooth eruption in CCD.

Jensen & Kreiborg (7) reported that the primary dentition is fairly normal in CCD, except for a slightly delayed eruption, whereas the permanent dentition is characterized by severe anomalies in tooth formation, maturation and eruption. All but one, of 18 cases evaluated by these authors had

EMNEORD

Cleidocranial dysplasia; supernumerary teeth; osteoclasts; tooth eruption

supernumerary permanent teeth, and two subjects developed a nearly complete supernumerary dentition mesial to the first molars. The supernumerary teeth developed lingually and occlusally to the normal teeth, and their maturation was delayed about 4 years compared to the normal teeth. All subjects had disturbances of eruption in the permanent dentition; both in regions with and without supernumerary teeth. The authors suggested that the supernumerary teeth mesial to the first molars represent a more or less complete 3rd dentition; that supernumerary molars develop through a repeating mechanism of normal molar development; and that the supernumerary teeth develop from remnants of the dental lamina, which have not been dissolved at the expected time. They concluded that the intraosseous stage of permanent tooth eruption is hampered in at least two ways in CCD:

- 1) If a supernumerary tooth develops in a given region, it forms occlusally to the normal tooth at the time when the intraosseous stage of eruption of the normal tooth should start and, thus, hinders eruption of the normal tooth.
- 2) The resorption of alveolar bone and primary teeth is severely delayed, even in regions without supernumerary teeth, because of insufficient osteoclast activity.

Based on these biological principles, Jensen & Kreiborg (8) suggested the following interceptive treatment strategies in CCD to promote spontaneous tooth eruption:

- 1) Early diagnosis of supernumerary teeth; supernumerary incisors can be expected to be diagnosed radiographically from 5-6 years of age and canines and premolars a couple of years later.
- 2) In regions with supernumerary teeth: removal of primary teeth, supernumerary teeth and overlying bone at the time when the roots of the normal teeth have reached about half of their final length.
- 3) In regions without supernumerary teeth: removal of primary teeth and overlying bone at the time when the roots of the normal teeth have reached about half of their final length.
- 4) Orthodontic treatment; if necessary, combined with autotransplantation of teeth, dental implants and orthognathic surgery.

This case report will illustrate the application of these treatment strategies and document the outcome of treatment.

Intraorale fotografier



Fig. 1. Intraoral photographs of the patient at ages A: 4.5 years; B: 10.2 years; C: 10.8 years; and D: 18.5 years.

Fig. 1. Intraorale fotografier af patienten ved aldrene A: 4,5 år; B: 10,2 år; C: 10,8 år; og D: 18,5 år.

Case study

A boy with CCD was followed from infancy till adulthood. He is the son of a woman with CCD who had previously been treated by us. CCD was observed in three generations, and all affected family members have a heterozygous deletion in the *RUNX2* gene (c.1121delG). The boy was diagnosed at birth because of severely delayed ossification of the calvaria and hypoplastic clavicles. He was followed clinically from 5 months of age to the age of 19 years. From 4.5 years of age the examinations were supplemented with annual intraoral photographs (Fig. 1), orthopantomographic x-rays (Fig. 2) and cephalometric x-rays (Fig. 3) to monitor dental and craniofacial development. This report will focus on the dental development.

Dental development

Primary dentition – The lower incisors started to erupt at 7 months of age. By 1.7 years all incisors had erupted and the canines had penetrated the oral mucosa. The second molars were relatively late to show signs of eruption, but at 4.5 years all molars were fully erupted and occlusion was normal (Fig. 1A).

Permanent dentition – At 5.5 years all teeth could be observed in the panoramic x-ray. No supernumerary teeth were seen at this age. However, the dentition was 9-12 months delayed in maturity compared to normative data (Fig. 2A).

Two supernumerary lower incisors were diagnosed at 6.7 years, and at 7.5 years a supernumerary tooth developed in the upper incisor region. No treatment was carried out at this stage, because of delayed root formation of the normal incisors. All first molars erupted spontaneously, but with a delay of about 2.5 years (Fig. 2B). At 9.5 years, all lower primary incisors were extracted, and the supernumerary lower incisors and overlying bone covering the permanent incisors were removed. At 10.2 years of age, all

Ortopantomografiske optagelser

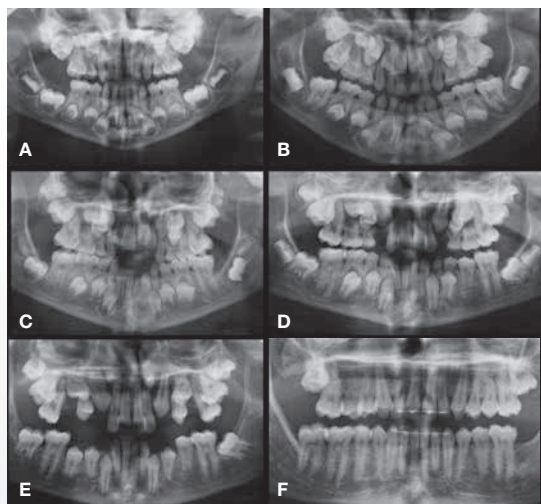


Fig. 2. Orthopantomographic films of the patient at ages A: 5.5 years; B: 8.5 years; C: 10.2 years; D: 12.2 years; E: 13.7 years; and F: 18.5 years.

Fig. 2. Ortopantomografiske optagelser af patienten ved aldrene A: 5,5 år; B: 8,5 år; C: 10,2 år; D: 12,2 år; E: 13,7 år; og F: 18,5 år.

upper primary incisors had been extracted; the supernumerary upper incisor had been removed, and the upper permanent incisors had been surgically exposed. The panoramic x-ray revealed that two supernumerary upper canines had developed. In addition, the upper second premolars were in an ectopic position, and the lower, left second molar had a mesioangular position. Dental maturation was, in general, delayed by 1½-2 years compared to normative data (Fig. 2C). The lower incisors were in spontaneous eruption at this stage (Fig. 1B). At 10.8 years, all permanent incisors were in spontaneous eruption (Fig. 1C).

About one year later, the upper primary canines were extracted, the supernumerary canines were removed and surgical exposure of the normal canines was carried out. At 12.2 years, the lower, left second premolar had erupted spontaneously, whereas none of the remaining teeth showed signs of spontaneous eruption although root formation of lower canines and all premolars had reached ½ to ¾ of their final root length (see Fig. 2D). All remaining primary teeth were extracted and the permanent teeth were surgically exposed. In addition, the lower third molars were removed and the lower, left second molar was surgically exposed. All canines and premolars erupted spontaneously except for the ectopic upper second premolars. Furthermore, the lower, left second molar and the upper second and third molars became impacted (Fig. 2E). The upper third molars and the lower left second molar were surgically removed. A fixed orthodontic appliance was used for a short pe-

Cefalometriske optagelser

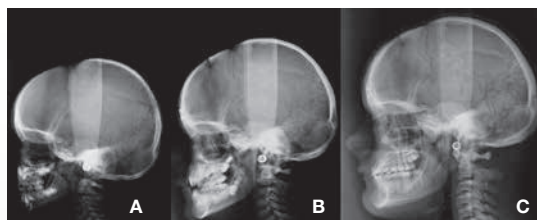


Fig. 3. Roentgencephalometric films in the lateral projection of the patient at ages A: 4.5 years; B: 11 years; and C: 18.5 years. Note delayed ossification of calvarial bones; numerous Wormian bones; hypoplasia of nasal bones and lack of development of a frontal sinus.

Fig. 3. Cefalometriske optagelser af patienten i lateral projekti-on ved aldrene A: 4,5 år; B: 11 år; og C: 18,5 år. Bemærk den forsinkede calcifikation af kalvarieknoglerne; talrige Wormske knogler; hypoplasia af næsebenene og manglende udvikling af sinus frontalis.

riod to open space for the upper second premolars; these teeth were surgically exposed and orthodontic traction was applied. The upper second molars remained impacted. The orthodontic treatment was completed with an acceptable aesthetic and functional result (Figs. 1D and 2F).

Discussion and conclusions

The primary dentition was normal and all teeth erupted spontaneously. Similar findings were previously reported by Jensen & Kreiborg (7). The reason for spontaneous eruption of primary teeth is probably that these teeth are only covered by a thin layer of bone; the need for osteoclastic activity in connection with eruption is thus minimal.

Five supernumerary permanent teeth were diagnosed in the incisor and upper canine regions. The teeth were positioned lingually and occlusally to the normal teeth and were delayed about 4 years in maturation compared to the normal permanent teeth. These findings are in agreement with previous observations by Jensen & Kreiborg (7). The normal teeth were also somewhat delayed in maturation, and this finding is in agreement with several previous studies (7,9,10). Tooth eruption was delayed both in regions with and without supernumerary teeth. This finding is also in agreement with previous observations (7). The supernumerary teeth obstruct the eruption path of the normal teeth which leads to arrested eruption, and diminished osteoclast activity is probably the cause of delayed tooth eruption in regions without supernumerary teeth (5,6). However, all first molars erupted spontaneously, although with a delay of about 2.5 years. Similar findings have previously been reported (7,9,10). The delay in eruption could mainly be explained by delayed maturation of these teeth. The fact that the teeth did



erupt spontaneously can probably be explained by the superficial position of these tooth buds in the jaws with limited bone coverage and lack of primary predecessors.

In general, the interceptive treatment strategies suggested by Jensen & Kreiborg (8), based on biological principles, were employed in the present case and lead to spontaneous eruption, although with delay, of all teeth mesial to the first molars, except for ectopic upper second premolars. However, these teeth responded well to orthodontic traction.

In conclusion, the treatment strategies used aimed at promoting spontaneous tooth eruption and were found to work well in the present case. These findings contradict the statement of D'Alessandro et al. (11) that natural eruption fails to occur in CCD.

Although our patient had several surgical procedures, these were all relatively minor. Furthermore, the corrective orthodontic treatment period was short since nearly all teeth erupted spontaneously. Thus, the burden of care would seem to be less than with the more aggressive corrective treatment approach-

CLINICAL RELEVANCE

The current case report supports the interceptive dental treatment strategies in cleidocranial dysplasia previously suggested to promote spontaneous eruption of permanent teeth. The strategies seem to result in a less

dramatic treatment for the patient and less complicated procedures for the dentist with a shorter total active treatment time compared to the corrective treatment strategies currently advocated in the literature.

es, currently advocated in the literature, with surgical exposure of nearly all permanent teeth and application of active orthodontic traction (9,10,12,13).

ABSTRACT (DANSK)

Cleidocranial dysplasi – interceptiv behandling af forstyrrelser af tandfrembrud

Baggrund – Formålet med den foreliggende rapport var at redegøre for behandlingsforløbet hos en dreng med cleidocranial dysplasia (CCD) behandlet i henhold til den interceptive strategi, der tidligere er blevet foreslået af Jensen og Kreiborg.

Patienttilfælde – Drengen blev fulgt fra den helt tidlige barndom til voksen alder med kliniske undersøgelser, fotografier samt ortopantomografiske og cefalometriske røntgenundersøgelser for at monitorere den kraniofaciale og dentale udvikling og behandlingsforløbet. Den primære dentition var normal. Den permanente dentition viste afvigelser i relation til såvel tanddannelse som tandmodenhed og eruption. Drengen udviklede fem overtallige tænder og havde alvorlige problemer med eruptionen af næsten

samtlig normale tænder. Det generelle behandlingsprincip var at fremme spontan eruption ved at fjerne overtallige tænder, primære tænder og knogle, der dækker de normale permanente tænder på det tidspunkt, hvor disse tænder har udviklet omkring halvdelen af deres endelige rod længde. Næsten alle tænder eruperede herefter spontant, og behovet for korrektiv ortodontisk behandling blev derved begrænset til en relativt kort periode.

Konklusion – Undersøgelsens resultater støtter forslaget om, at patienter med CCD kan behandles interceptivt ved fjernelse af hindringerne for normal tandruption (overtallige tænder, primære tænder og alveolær knogle) på det tidspunkt, hvor de normale permanente tænder har udviklet omkring halvdelen af deres endelige rod længde.

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