



CASE REPORT

Cleidocranial Dysplasia: A Case Report

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Abstract

Cleidocranial dysostosis (CCD) is a congenital disability that may affect bones and teeth. It is a rare disease that can be caused by inheritance or a genetic mutation, irrespective of ethnic group. Treatment of CCD aims to achieve optimum function and aesthetic results, and improve the patient's quality of life and self-confidence. In this case report, we present a nine-year-old male patient with CCD seeking treatment due to delayed eruption of teeth. Many other dental problems are also presented in this case, such as supernumerary teeth, retained deciduous teeth, malformed teeth and multiple impacted permanent teeth. The patient underwent both surgical and orthodontic procedures. Early diagnosis of CCD leads to optimum management, resulting in improvement of patient's quality of life.

Keywords: Cleidocranial dysplasia; Dysostosis; Dentofacial deviation; Supernumerary teeth; Clavicular defect.

Introduction

Cleidocranial dysostosis (CCD), also called cleidocranial dysplasia, is a congenital disability that may affect teeth and bones.¹ The clavicles are usually either poorly developed or absent in CCD, which makes it possible for the shoulders to be brought together closely.¹ It also causes delay of closure of the front of the skull, and the affected individuals are often shorter than average. Other features may include a prominent forehead, wide-set eyes, abnormal teeth, and a flat nose. Symptoms vary among people, however, intelligence is typically average. The condition can be either inherited or genetic.² The mandible in CCD can be prognathic and the maxilla and other facial bones are hypoplastic. There can be a failure of closure of

fontanel, resulting in a soft spot on the head.³ One of the significant findings of CCD is having multiple supernumerary teeth. Other features of CCD include failure of eruption of permanent teeth, which may be due to deficient cementum formation. Other general characteristics are hypertelorism, short middle fifth phalanges, vertebral abnormalities, scoliosis, spina bifida, and syringomyelia.^{4,5}

The management of CCD involves a multi-disciplinary team consisting of mainly dentists, orthodontists and maxillofacial surgeons. Following improvement of dental health by dentists, combined orthodontic and surgical treatment aimed at extraction of supernumerary teeth as well as disimpaction and alignment of unerupted teeth is carried out.

Case Report

A nine-year-old Bahraini boy attended the Dental & Maxillofacial clinic at the Bahrain Defense Force Hospital complaining of missing teeth. His medical history was normal. His parents confirmed that he was born normal with good health. The family did not recall any inherited problems or a similar condition among their relatives. The child had undergone no prior treatment and this was his first visit to the Bahrain Defense Force Hospital.

A general physical examination revealed a short-statured, well-oriented young boy with narrow, drooping shoulders. Facial examination showed a brachycephalic head with frontal and parental bossing, hypoplastic maxillary and zygomatic bones, hypertelorism, bulging calvarium, and depressed nasal bridge with a broad alar base (Figures 1 and 2).



Figure 1: Anterior view of the patient showing the narrow drooping clavicles.



Figure 2: Lateral view of the patient showing bulging calvarium and depressed nasal bone.

The intra-oral examination revealed multiple missing teeth and multiple carious deciduous teeth (Figure 3). It was also noticed that the patient had swelling in the lower mandible lingually.



Figure 3: Anterior intraoral image showing multiple clinically missing teeth, and carious teeth.

Special investigations such as Cone Beam Computerized Tomography (CBCT) revealed multiple dental problems including retained and impacted teeth in the maxilla and mandible. The presence of supernumerary teeth and underdeveloped maxillary sinus were noted as well (Figure 4).



Figure 4: A scan section of CBCT showing multiple retained and impacted teeth in maxilla and mandible, supernumerary teeth and underdeveloped maxillary sinus.

Based on the above signs and symptoms, the patient was diagnosed with cleidocranial dysplasia and the lingual swelling was diagnosed as a ranula.

The general management of such conditions is multidisciplinary, involving general dentistry, orthodontics, maxillofacial surgery, and prosthodontics.

The management of the case started with oral hygiene improvement, followed by extraction of badly broken-down teeth and restorations of the

carious lesions. After the stabilization stage, the patient underwent the care of a multidisciplinary team comprising of oral and maxillofacial surgeon, paedodontist, periodontist and orthodontist. He underwent a surgical procedure under general anesthesia for extraction of multiple supernumerary teeth, exposure of the impacted teeth and excision of the ranula.

The patient is still undergoing treatment. The ongoing treatment has currently resulted in eruption of some of the previously impacted permanent teeth, and we further aim to guide the eruption of the remaining impacted teeth, to achieve a satisfactory result.

Discussion

Sneha et al., reported that CCD cases revealed the absence of clavicles, open skull sutures, unerupted permanent successors, and supernumerary teeth during radiological examination, similar to findings in the present case.⁹

In a general physical examination, Sarbjeet Singh et al., and Kolokitha et al., reported cases with short-stature and narrow, drooping shoulders. Both of them also noted a brachycephalic head with frontal and parietal bossing, hypoplastic maxillary and zygomatic bones, bulging calvarium and sunken nasal bridge.^{10, 11} All these features were also noted in this case.

In both intraoral and radiographic examinations, Gülay et al., and Teresa et al., noticed the presence of an excessive number of supernumerary teeth, whether in the maxilla or the mandible, among CCD cases.^{12,13} The supernumeraries were also present in the present case. Das Gupta R, et al., and Yanfei et al., reported that the major features of CCD included aplastic or hypoplastic clavicles, delayed closure of the sagittal fontanel, failure of permanent teeth eruption, and multiple supernumerary teeth.^{14,15}

All of these findings, pathognomonic for the diagnosis of CCD, were noted in this case.

In a similar approach to management as in this case, Yanfei et al., went through a process wherein they first extracted all the primary teeth as well as the supernumerary teeth that affected the eruption of

the permanent teeth. Then, the impacted permanent teeth were surgically exposed and followed up with the orthodontist, to control the eruption of the teeth by appliances.¹⁵

They reported that surgical exposure combined with orthodontic traction is an effective treatment for patients with CCD. The success of the treatment depends on many factors such as the age of the patient, compliance, economic circumstances, and the condition of permanent dentition.¹⁵

Achieving an accurate diagnosis relies on early detection of the clinical features of CCD and leads to an effective treatment plan.

Management of CCD may require a lengthy treatment process. Both the patient and the parents need to be made aware of the length and stages of treatment that would require multi-disciplinary team care.

The present case is still undergoing treatment, with results promisingly aimed to improve aesthetics, oral function, psychosocial wellbeing, and quality of life.

This case report highlights the importance of multidisciplinary treatment planning in achieving the best result possible for the patient, and it confirms the role of each specialty in providing the best outcome. Publishing a case report such as this is beneficial and can be used in the future as a reference for related cases and as an educational source for practitioners, patients, and even their families. By comparing initial manifestations and post treatment results, the management plan can be used as a reference for future patients, who may be hesitant in going through such lengthy and possibly invasive procedures.

Conclusion

Many patients with cleidocranial dysostosis have only limited number of findings and are often not recognized to have the disease. Therefore, the diagnosis may be missed, delayed or detected incidentally. Family history, excessive mobility of shoulders, and radiographic pathognomonic findings of the chest, jaws, and skull are helpful tools in confirming the diagnosis.

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