

A Case of Cleidocranial Dysostosis: Dilemma for a Prosthodontist

Shuchi Tripathi · Raghuwar D. Singh ·
Saumyendra V. Singh · Pooran Chand

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Abstract Cleidocranial dysostosis (CCD) is an uncommon, generalized skeletal disorder characterized by delayed ossification of the skull, aplastic or hypoplastic clavicles, and complex dental abnormalities such as retention of multiple deciduous teeth, impaction or delayed eruption of permanent teeth and presence of supernumerary teeth. This case report describes a 30-year old male patient of CCD with classical findings and the challenges faced in his prosthodontic rehabilitation. Conventional complete dentures following interim complete dentures therapy had to be given as all other treatment modalities were ruled out because of anatomic limitations. Despite the disadvantages of removable dentures the patient adapted admirably to them with significant improvement in self-esteem. Though orthodontic and surgical correction may be the treatment of choice, the need to preserve the alveolar ridge could make fabrication of removable dentures in such patients, a viable option.

Keywords CBFA1 gene · Cleidocranial dysostosis · Complete dentures · Impacted teeth · Wormian bones

Introduction

Cleidocranial dysostosis (CCD) (cleido—collar bone, cranial—head, dysostosis—abnormal forming of bone) is a rare congenital defect of autosomal dominant inheritance primarily affecting bones that undergo intra-membranous ossification. It is also known as Marie and Sinton disease, mutational dysostosis, craniocleido dysostosis, cleidocranial dysplasia and osteodental dysplasia [1, 2]. About 500 cases have been reported world-wide [3]. Only few have been reported in India [4].

CCD is caused by changes in the CBFA1 gene, which is a DNA transcription factor for osteoblast-specific genes [5]. This gene is located on chromosome 6, which encodes a protein necessary for the correct functioning of osteoblast cells. However 40 % of cases of CCD appear spontaneously with no apparent genetic cause [6]. This condition is of clinical significance to every dentist due to the involvement of the facial bones, altered eruption patterns and multiple supernumerary teeth [7].

Case Description and Results

A 30-year old male patient came to the Dental Faculty of the Institute with the chief complaint of pus discharge in the maxillary posterior region and difficulty in chewing. On inquiring about his dental history, the patient revealed that several of his teeth had never erupted and he had been to a dentist several times in order to have his mobile teeth extracted. Though no relevant family history was found, the general and facial examination of the patient revealed a short stature, the ability to voluntarily touch his shoulders together in front of his body (Fig. 1), an enlarged forehead with frontal bossing, slurred speech, brachycephaly,

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S. Tripathi (✉) · R. D. Singh · S. V. Singh · P. Chand
Department of Prosthodontics, Faculty of Dental Sciences,
C.S.M. Medical University (Upgraded K.G.M.C.),
C-13, Arawali Marg, Indiranagar, Lucknow 226016,
Uttar Pradesh, India
e-mail: dr08shuchi@yahoo.co.in



Fig. 1 View of the patient's shoulders in contact

midfacial hypoplasia with low nasal bridge, hypertelorism and mandibular prognathism. Skeletal X-rays of the patient were made which showed undergrowth of the clavicle and scapula, wormian bones and calvarial thickening.

The intra oral examination revealed presence of abnormally retained deciduous teeth 53, 63 and 71 and permanent teeth 17, 16, 11, 26, 27, 37, 36, 35, 34, 41, 46 and 47, a high arched palate, enamel hypoplasia, variable size and shape of erupted teeth and third grade mobility of all existing teeth. The full mouth series of radiograph including the orthopantamograph showed abnormally impacted permanent dentition, multiple supernumerary teeth and root dilacerations (Fig. 2).

Due to the combined factors of mobility of all existing teeth, age of patient and lack of anchorage for orthodontic correction, it was decided that conventional complete dentures would have to be fabricated. It was also decided that the retained impacted teeth would not be removed. Interim complete dentures were first fabricated and inserted

immediately after extraction of all teeth thus maintaining pre-extraction vertical dimension. Follow-up was done and after 3 months, conventional complete dentures fabrication was initiated.

The maxillary and mandibular ridges were found to be irregular and asymmetric, with the maxillary arch being tapered and having abnormally bulging horizontal vaults with sharply furrowed mid line raphe (Fig. 3). Preliminary impressions of maxillary and mandibular ridges were made with irreversible hydrocolloid (Zelgan, Dentsply India Pvt. Ltd.) and poured. Relief was provided at the previously marked areas on the cast where impacted teeth were present and in mid-palatal raphe region with double spacer wax (1 mm) before fabricating the custom trays. Conventional border molding was done with low-fusing impression compound (DPI Tracing sticks, Dental products of India, Mumbai, India), final impressions were made with zinc-oxide eugenol impression paste (DPI Impression paste, Dental products of India, Mumbai, India) after removing the spacer wax and by creating relief holes in the mid palatine raphe and horizontal shelves in the maxilla and in the anterior region of the mandible where impacted teeth were present. Jaw relation was recorded, while maintaining the patient's original vertical dimension utilizing the interim complete denture as guideline followed by teeth arrangement (Cosmo HXL, Dentsply Ltd., Surrey, UK). During teeth arrangement balanced occlusion was established on Hanau Wide-View articulator and the arrangement was verified esthetically and functionally. Dentures were cured, corrected occlusally and inserted. Instructions were given regarding denture use and emphasis was given for regular follow-up appointments. Oral pantomographs were repeated after every 3 months to verify the condition and eruption, if any, of the impacted teeth. The patient was satisfied with the esthetic and functional outcome of the denture (Fig. 4). Marked psychological build up of the patient was seen with increase in confidence level and fluency in speech.



Fig. 2 Pre-operative ortho pantamogram



Fig. 3 Maxillary ridge after post extraction healing



Fig. 4 Post-treatment *frontal view* of the patient

Discussion

Dental problems are often the most significant complications of CCD patients and therefore appropriate dental work is vital. Orthodontic with surgical correction is generally the treatment of choice [8–10], but extreme mobility of the teeth, increased age and anchorage loss were the main hindrance in orthodontic treatment [11]. The process of involving orthodontics, orthognathic surgical interventions and interim prosthesis would take several years until patient will receive definitive prosthesis [12]. Implant placement was also considered but due to unavailability of proper bone and presence of impacted teeth this treatment modality was not possible.

Delayed eruptions of permanent teeth have been noted due to absence or paucity of cellular cementum on the roots of the teeth [13]. Some authors suggest that the removal of primary or supernumerary teeth do not promote eruption of impacted permanent teeth and in addition, permanent teeth may be difficult to extract due to malformed roots [3]. Rehabilitation of the present case followed the same principle. The embedded teeth may serve not only to maintain the alveolar ridge but the multiple surgeries needed for extracting these teeth would have led to much bone loss and psychological trauma to the patient.

The maxillary arch was tapered with bulged palatal shelves and deeply furrowed mid-palatine raphe. This adverse condition predisposes to less stability, retention and support in the denture. Therefore modifications were made at every steps of denture construction. The primary impressions were made with irreversible hydrocolloid to exert less pressure over the denture bearing area and to record accurately the undercuts and irregularities present to improve retention. Similarly emphasis was given on

applying low pressure over concerned areas (as described previously) during final impression procedure. The pre-extraction records of the natural teeth served as a guide for fabrication of interim complete dentures at the normal vertical dimension, which was maintained during conventional complete denture fabrication. It is noticeable that the mandibular prognathism was due to prominent lower two-thirds of the body of the mandible. Upper one-thirds containing alveolus of the mandible was in end-on relation with maxilla which was converted into class 1 relation, thus providing more natural appearance. Bilateral balanced occlusal scheme using 20° of cuspal angulation was provided to ensure denture stability and support.

As impacted teeth were not extracted, regular follow-ups were essential to monitor the patient's condition. The patient was examined clinically and the locations of impacted teeth were examined by orthopantomogram after every 3 months for 1 year. According to the American College of Radiology (ACR) an orthopantomogram involves minimal radiation to the tune of 0.001 mSv, which is less than the radiation an individual receives during the course of an average day [14]. Despite the minimal risk involved the patient was informed of the ill effects of radiation and his consent taken along with obtaining ethical approval. This was because the benefits of frequent OPGs outweighed the risks in this patient. The patient's psychological status was significantly improved as evaluated using the quality of life scale (Likert's scale) [15] and speech was evaluated by a speech therapist resulting a definite improvement in the post rehabilitation phase of the patient.

The existing disharmonies severely limited the available treatment modalities, due to which a 30-year old young man had to wear complete dentures. However the purpose of maintenance of function as well as esthetics could be solved with satisfactory result in the present case report.

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