

TREATMENT APPROACHES TO CLEIDOCRANIAL DYSOSTOSIS: A REVIEW AND A CASE REPORT

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ABSTRACT:

Cleidocranial dysostosis is a generalized skeletal dysplastic condition and primarily affects the development of the bones and teeth. The genetic etiology lies on chromosome number 6p21, core binding factor (CBFA1). This paper explains the treatment procedure carried out for a sixteen year old female patient diagnosed with cleidocranial dysostosis. Therapy includes surgical removal or exposure of impacted teeth, extraction of supernumerary teeth along with orthodontic treatment.

Key words: Cledio-cranial dysplasia, supernumerary teeth, diagnosis

INTRODUCTION

Cleidocranial dysostosis (CCD) is a generalized skeletal dysplastic condition and affects the development of the bones and teeth. It has a low prevalence of 1 per 10,000 population. CCD is under diagnosed as the medical complications involved in it are less as compared to other skeletal dysplasias.^{1,2}

The typical findings of this disorder include a variety of skeletal abnormalities and multiple oral manifestations like aplasia of the clavicle, short height, delayed and abnormal cranium ossification.³ The cleidocranial dysostosis patients appear to have a peculiar appearance of bulky forehead, hypertelorism, and midfacial hypoplasia.

At a very early stage it was thought that this disorder involved only intramembranous bones, hence the term 'cleidocranial' as coined by Marie and Sainton in 1897.⁴ Hesse⁵ has described the anomalies of teeth and jaws connected with cleidocranial dysostosis in detail.

The gene has been identified on the short arm of chromosome number 6p21, core binding factor (CBFA1). The literature suggests that around 20-40% of cases represent new mutations in the transcription factor. The CBFA1 gene regulates the formation of osteoblasts which is absolutely necessary for both membranous and endochondral bone formation, and can be linked to delayed ossification of the bones and dentition as seen in this syndrome.⁶ However, 40% of these cases appear spontaneously with no genetic etiopathogenesis.⁶

There are few genotype phenotype studies⁷ which show that skeletal and dental development could be related to the type of mutation in the Runt-related transcription factor

2(RUNX2) gene. It is a gene which encodes a protein which is necessary for correct functioning of osteoblast cells

As compared to the cranium the face appears to be small relatively because maxillary, lachrymal, nasal, and zygomatic bones are hypoplastic. The mandible appears to be protrusive as the maxilla is underdeveloped. Dental abnormalities involve delayed eruption of permanent teeth, multiple supernumerary teeth, retained primary teeth, dilaceration of teeth, and reduced alveolar height, crown and root abnormalities, crypt formation around impacted teeth, and a high palate⁸

This paper describes the treatment done for a sixteen year old female patient diagnosed with Cleidocranial dysostosis and its outcome.

CASE REPORT

A sixteen year-old female patient with CCD reported to our department with chief complaint of permanent teeth not erupting causing disability in eating, poor dental aesthetics and facial appearance. There was no family history of any such disorder and no parental consanguinity was reported. Figure 1 shows a straight profile, frontal bossing and depressed suborbital regions in extraoral pictures of the patient.



Figure 1 Pretreatment Extraoral Pictures

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On performing oral examination, a mixed dentition with the following dental formula and an Angle's Class III molar relationship was observed. (Figure 2)

7 6 E D C A	A B C D E 5 6 7
7 6 5 4 D 3 2 1	1 3 4 C E 5 6 7

Oral evaluation also revealed missing both maxillary and mandibular permanent teeth (Figures 2) Carious and fractured primary maxillary incisors and right mandibular first molar.



Figure 2 Pretreatment Intraoral photographs

The pretreatment panoramic radiograph (figure 3) showed presence of impacted and supernumerary teeth in both upper and lower arches. The following teeth can be viewed in the OPG:

7 6 E D III C A I	I A B C III E 5 6 7
8 7 6 5 4 D IV III 3 2 1 I	1 3 4 C E 5 6 7 8

Teeth in roman numbers represent the supernumerary teeth.

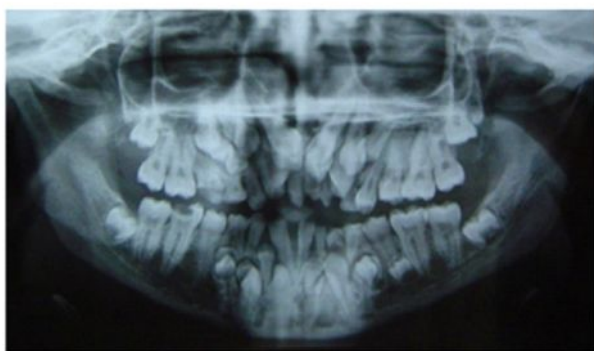


Figure 3 Pretreatment OPG

The clinical findings were confirmed by a lateral cephalogram, skeletal class III malocclusion with ANB of -4 degree. The mandibular plane angle was increased, showing vertical growth pattern. Cervical vertebrae shows deep concavities at the lower borders of C2, C3 revealing almost completion of growth. (Figures 4)

The diagnosis was formulated based on the following findings, bilateral hypoplasia of the clavicles, an enlarged cranium, frontal bossing, depressed suborbital region, failure of eruption, and multiple supernumerary teeth. This patient also had a short height and a history of retarded growth. However, cognitive development was entirely within normal limits. The craniofacial findings

included delayed closure of cranial fontanels and suture and brachycephalia. Therefore, the patient was diagnosed with skeletal class III mandibular prognathism and maxillary retrognathism accompanied by multiple congenitally impacted and supernumerary teeth.



Figure 4 Pretreatment Lateral cephalogram

Literature included a variety of management approaches for treatment of CCD. In this case report, we followed the Belfast-Hamberg approach which is a single step procedure involving removal of all primary and supernumerary teeth. These extractions were followed by bonding of all permanent teeth with Standard Edgewise appliance.022" slot and bonding of impacted teeth to extrude teeth into the oral cavity. A transpalatal arch was given with soldered hooks used to extrude impacted teeth in the mandibular arch.

Midtreatment pictures (figure 6) show all permanent teeth aligned in one arch with anterior open bite which was corrected by using box elastics.

DISCUSSION

The management of CCD is a multidisciplinary approach involving orthodontics, orthognathic surgical interventions, and interim prostheses. This process of involving all these disciplines can take several years until patients can receive their definitive prostheses.

Cleidocranial dysplasia manifests with many anomalies of the teeth and jaws. Depending on each anomaly, various authors have prescribed specific treatment for each as described in table 1.

Later four distinct approaches were introduced depending on case to case. The four approaches were the Toronto-Melbourne approach⁹, Jerusalem approach^{9, 10}, Belfast-Hamburg approach^{9, 10} and Bronx approach¹⁰.



Figure 6 Midtreatment intraoral pictures

After 18 months of orthodontic treatment and composite build-up of 22(Lateral incisor of second quadrant) the case was functionally and aesthetically completed. (Figure 7)



Figure 7 Post treatment intraoral pictures

As described in table 2, several procedures at different ages are involved in Toronto-Melbourne. Jerusalem approach has divided treatment in two phases. Whereas Bronx approach involves a two phase or maximum 3 phase procedure unlike Belfast- Hamburg approach which is a single step procedure.

Treatment plan proposed for our case was very similar to the Belfast- Hamburg approach as described in the table 2.

Table 1 Cleidocranial dysplasia: orodental anomalies and management options

Anomaly	Management option	Rationale
Retained deciduous teeth	Removal ^{11,12,13,26}	Assist eruption of permanent teeth
Supernumerary teeth	Removal ^{11,12,13,18,26}	Assist eruption of permanent teeth
Permanent teeth abnormalities	Removal ^{16,20,22,23,26,28}	Removable full and partial dentures
	Retention ^{25,27}	Abutments for fixed appliances
Unerrupted teeth	Surgical exposure ^{12,13,17}	Support for overdenture
	Orthodontic eruption ^{9,10,13,14}	Functions, aesthetics and alignment
	Implants ^{37,26}	Support overdenture
		Guide impacted teeth into occlusion
	Autotransplantation and/or surgical translocation ^{9,10,14,19,24}	Function and esthetics
Malocclusion	Removable/Fixed orthodontic appliances ^{9,11,20}	Function and esthetics
Palatal vault narrow-high arched	Expansion using removable orthopedic appliance ²⁶	Reduce crowding

Although Toronto-Melbourne approach offers a series of several extensive and minor surgical procedures, over a long period. Partial success in encouraging spontaneous eruption in this condition would be an optimistic forecast. In Jerusalem approach, it causes considerable delay before artificial enhancement of eruption is initiated. But Belfast-Hamburg offers a single, all-encompassing surgical procedure to eliminate the superfluous (deciduous and supernumerary) teeth and to expose the remainder at the age of 12-14 years.

Table 2- Cleidocranial dysplasia: 4 distinct management approaches

Approach	Procedure
Toronto-Melbourne ⁹	
Several procedures	
Age: 5-6	Extraction of primary anterior teeth
Age: 6-7	Primary incisors are exposed and healing is allowed
	Orthodontic brackets are bonded to permanent incisors
	Extraction of primary posterior teeth
Age: 9-10	Surgical exposure of permanent bicuspid
	Surgical removal of supernumerary teeth and healing allowed
Age: 9-12	Placement of orthodontic brackets on permanent canines and bicuspid teeth
Jerusalem ^{9,10}	
Age: 10-12 Phase 1:	Extraction of anterior primary teeth
	Extraction of all supernumerary teeth
	Surgical exposure of permanent incisors
	Orthodontic attachments are bonded to permanent incisors
	Surgical flaps are closed
Age: 13 and older Phase 2:	Extraction of posterior primary teeth
	Surgical exposure of unerupted canines and premolars of permanent dentition.
	Bonding of orthodontic attachments
	Surgical flaps are closed
Belfast-Hamburg ^{9,10}	Removal of all primary and supernumerary teeth
Single procedure	
Age: not specified	Surgical exposure of all impacted teeth
	Surgical packs are placed (to prevent healing of bone and soft tissue over teeth)
	Healing by secondary intention
	Bonding of orthodontic attachments
	Placement of orthodontic appliances on fully erupted dentition
	Elastic thread is placed between brackets on impacted teeth and the archwires
Bronx ¹⁰	Phase 1:
Two at most three Procedures	Removal of all primary and supernumerary teeth
Age: not specified	Surgical flaps are closed
	Phase 2:
	Unerrupted permanent teeth are exposed Orthodontic brackets are placed Surgical flaps are closed and overdenture is placed Conventional orthodontic appliances are placed
	Phase 3:
	Leforte osteotomy-orthognathic surgery
	Dental implants are placed

CONCLUSION

Cleidocranial dysostosis shows variable expressivity but early diagnosis through oral findings is possible. In addition to the oral evaluation, diagnosis of this rare syndrome requires a thorough skeletal evaluation. Achieving a well-functioning permanent dentition and an aesthetically satisfying facial appearance should be the main objectives of treatment of this rare clinical entity.

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