# CLEIDO CRANIAL DYSTOSIS: PROSTHODONTIC CHALLENGES AND REHABILITATION

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## ABSTRACT

This clinical report discusses the interdisciplinary management of a young Indian girl with a non familial history of unique and rare syndrome cleido cranial dystosis. The case report highlights the Prosthodontic challenges and objectives in rehabilitating the clinical situation. The author describes fabrication of removable overlay prosthesis over short copings on remaining natural teeth as a definitive treatment. The report highlights the importance of team work and Prosthodontic therapy in esthetic, functional and psychosocial rehabilitation of the patient.



## KEYWORDS

cleido cranial; aplasia; hypoplasia; clavicle; dentofacial


## INTRODUCTION

Cleidocranial Dysostosis is a congenital craniofacial disorder affecting skeletal and dental system. It is characterized by a triad consisting of; Clavicular aplasia or hypoplasia delayed ossification of the fontanel's and sutures of the vault of the skull and hereditary transmission1. Salzmann, describes Cleidocranial Dystosis as a disease of bone growth which occur with different degree of intensity2. It is commonly known as Marie- Sainton syndrome after the duo coined the descriptive title cleidocranial dysostosis in (1898). Later Hesse (1926) established the association of dentition and jaw findings with cleidocranial dysostosis3. Jackson in (1951) called it as osteodental dysplasia4.

It has a genetic autosomal pattern of inheritance with equal predilection for both the sexes5. The gene for cleidocranial dysostosis has been mapped on the short arm of chromosome 6p21, core binding factor alpha-1 (CBFA1). This disorder can be caused by mutation in the transcription factor CBFA1 (RUNX2). (RUNX 2) is the master gene in formation of bone & dental tissue6. The most common findings are absent or rudimentary clavicles with approximation of shoulders in the midline7. Retarded development of bones of the face and jaws, enlargement of lateral skull form and delayed closure of skull sutures. Skull - large number of wormian bones with supernumerary centers of ossification8. Extra Oral signs may include; Absence of nasal bone, frontal bossing, prominent chin and marked maxillary hypoplasia giving it a Quarter moon Physiognomy described by Hultkranz (1908).

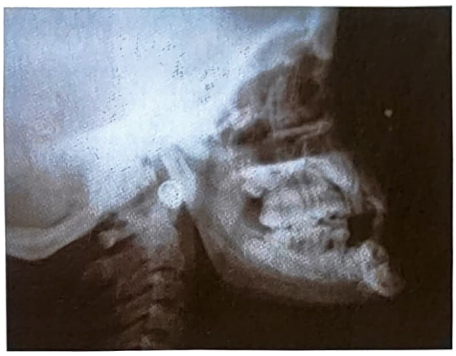

FIG 1: Pre-operative Lateral Cephalogram

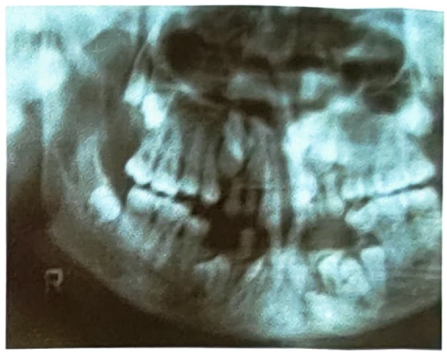

FIG 2: Pre-operative OPG showing multiple impacted supernumerary teeth

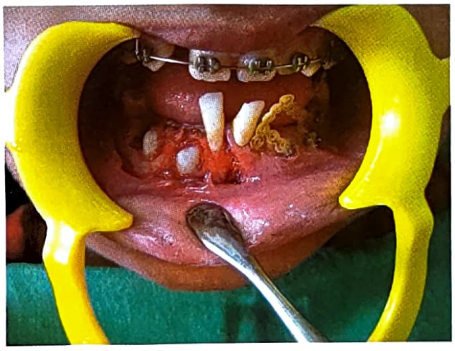

FIG 3: Surgical Exposure of Impacted Teeth

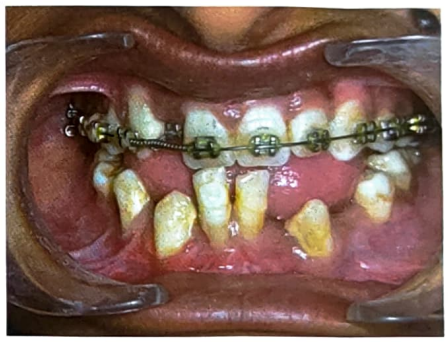

FIG 4: Orthodontic alignment of maxillary teeth and post operative mandible (Intraoral view)

Dental findings may include delayed eruption of deciduous dentition. Multiple impacted deciduous and permanent teeth. Presence of multiple supernumerary teeth particularly in the maxillary anterior and mandibular premolar region. Abnormal large spacing in the lower incisor area due to a wide alveolar bone and the presence of second molar sign- eruption of the second molars despite persisting primary dentition9.

Early diagnosis should be established so as to avoid prolonging aesthetic & emotional problems and to enable planning for what is a long period of treatment10. Differential diagnosis of delayed eruption and multiple supernumerary teeth must be established. Molecular genetic analysis for early detection of CCD should also be insinuated11. Establishing diagnosis of CCD may be challenging due to difficulties in identifying the genetic& developmental characteristics. Also the tendency for spontaneous occurrences and a wide range of Clinical variability ranges from unrecognizable to full blown cases12.

Orthopantomographs and lateral cephalograms may be suggestive of CCD and should be advised on first indication. Classical Panoramic signs may include; conglomeration of retained deciduous

teeth, only few erupted permanent teeth, multiple supernumerary and majority unerupted permanent deeply embedded teeths. The shape of the ascending ramus is characteristic (Mc Namara) with near parallel-sided borders, slender pointed coronoid process (upward, posterior direction) sigmoid notch often had a deep U-shaped curve. Lateral cephalogram may reveal Kyphotic sphenoid bone Missing nasal bone, wormian bone, deficient gonion angle, small maxilla especially in its vertical dimension. The articular eminence is often flat and zygomatic arch may be thin and may have a characteristic downward bend13.

Patients with CCD may manifest in variety of dentofacial needs requiring an interdisciplinary care involving Orthodontics, Maxillofacial surgery, Prosthodontics and Psychologist. Once diagnosed, team approach may be implicated for a successful clinical and social Rehabilitation. The clinical concerns may require immediate addressing of issues related to vertical dimension, depressed maxilla and improvised esthetic and functional needs of the patient14.

This article discusses about a case of CCD in a young girl living in rural north Indian population. The case discusses the role of team approach and innovative prosthodontic approach in rehabilitating the situation.

## CLINICAL REPORT

A 16 year old girl reported to orthodontic clinic with chief complaint of few missing lower anterior teeth and abnormally inclined lower central incisors. Patient complained of poor esthetics and requested prosthodontic rehabilitation as she wished to get married. Clinical examination revealed over retained maxillary deciduous lateral incisors and canines bilaterally (52, 53, 62, 63) and mandibular (83, 75).

Besides over retained deciduous numerous supernumerary and impacted teeth were also present. The maxillary lateral incisors and canines were impacted bilaterally with one supernumerary in the right canine region and two in the incisor canine region on the left side. In the lower arch the lateral incisors, canines and first premolars were impacted bilaterally (32, 33, 34, 42, 43, 44). The left lower second premolar was also impacted. In the mandibular arch, supernumeraries were seen in right canine, premolar region and two in the left premolar, one in canine region and one in incisor region of the left side.

All the deciduous teeths were extracted and in the maxilla supernumeraries were extracted and impacted teeth were bonded with .022 MBT mechanotherapy. We could get all the impacted teeth in place in the upper arch. In the lower arch the impacted teeth were extracted. In the lower arch corticotomy was performed to expose 44, 35 and rest all other impacted and supernumerary teeths were extracted.

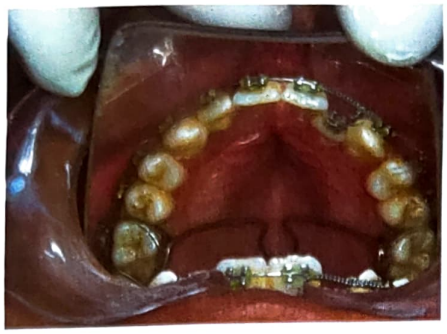

FIG 5: Orthodontic treatment of maxillary arch (occlusal view)

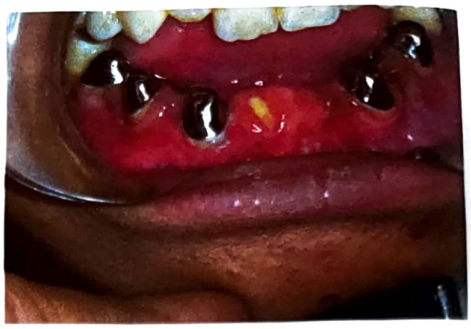

FIG 6: Short Ni-Cr copings in position

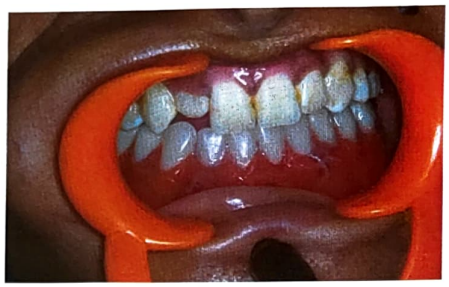

FIG 7: Final Prosthesis in position

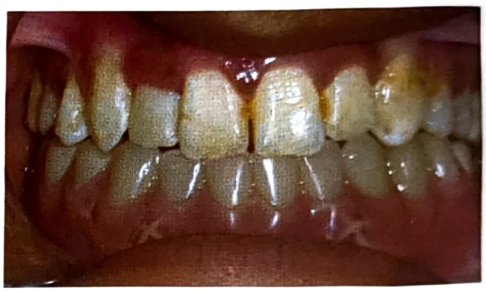

FIG 8: Post-operative extra oral view

OPG showed a long and slender ramus and impacted supernumerary distomolar in the upper right region. The second molar sign was there with two erupted lower incisors which were periodontally compromised. Since patient refused extraction of her lower central they were submerged in the bone after resection of supraosseous portion. The chest x ray revealed absence of the clavicle and was substantial in establishing the diagnosis.

The Prosthodontic management was complex and involved rehabilitation of poor esthetics, function and psychosocial needs of the patient. An interim all acrylic (Lucitone) prosthesis was fabricated to provisionally address her esthetic needs and it also gave an opportunity to access her intermaxillary relation and the type of final prosthesis that would best restore her to optimum. The patient wore the interim prosthesis for one year and reported no discomfort except for few initial adjustments.

It was decided to accept the patient existing occlusal plane as the definitive plane. Also, since the maxillary teeth were orthodontically aligned ,no prosthetic concerns were expressed in maxilla except the inadequate length of maxillary right lateral incisor which was decided to be restored with direct composite resin veneer. In the mandibular arch 33, 35,43,44,45 were endodontically treated and prepared with chamfer finish line to receive short copings for the fabrication of overlay prosthesis.

Copings were fabricated in Ni-Cr alloy and were cemented with multilink resin cement to ensure good metal bonding. Subsequently, Final impressions were made in a border molded custom tray in medium body ( Dentsply ) elastomeric material for the fabrication of metal framework of the overlay prosthesis. A cast metal (Co-Cr) framework was fabricated and tried in patient mouth for fit and stability. Jaw relations were made in pink hard wax ( DPI ) and semi-anatomic tooth form (Cosmo) were selected for occlusion with opposing natural teeth in centric occlusion. The overlay denture was processed in Heat cure acrylic resin and delivered to the patient after adjusting the occlusion.

There was a remarkable improvement in the esthetics with correction of intermaxillary relation. Patient was satisfactorily rehabilitated and confirmed efficient chewing ,speech and elevated social self esteem.

## DISCUSSION

This paper discusses the rehabilitation of a

rare craniofacial syndrome which is genetically expressed and is congenital disorder affecting skeletal & dental system. Because of the rarity of the condition, guidelines for the treatment of the CCD are rather sparse in the literature. In this report a interdisciplinary approach involving surgical, orthodontics and Prosthodontic treatments were required to successfully rehabilitate the patient.

The patient may exhibit variety of signs and it is important to identify early signs besides molecular genetic analysis so as to initiate rehabilitative measures. The concerned patient presented with multiple challenges including esthetic, functional and psychosocial. It was realized that the patient reported late for the treatment and was under social pressure. Surgical removal of impacted and multiple supernumerary teeth were planned so as to allow orthodontic alignment of the remaining teeths. Concomitant to orthodontic therapy further analysis was done to address patient functional and residual esthetic demands.

The prosthetic goals were established after completion of the orthodontic therapy and were aimed towards replacement of missing teeths, and correction of abnormal three dimensional intermaxillary relationships for the best possible profile and functional position. The selection of prosthesis type for the patient is best governed by the remaining natural teeth ,their position and existing intermaxillary relationship. Since the maxilla was restored orthodontically , the remaining natural teeth in the mandibular arch were prepared to serve as overlay copings for the fabrication of overlay prosthesis.

The overlay prosthesis gave an opportunity to utilize natural teeth support, maintained proprioception, preserved the natural masticatory force and allowed correction of the deficient mid face . The prosthesis being removable also allowed the patient ease of cleansibity and a non-invasive nature of prosthodontic rehabilitation.

## SUMMARY

This case report describes the interdisciplinary management of a young female diagnosed with CCD using undermentioned key points for achieving optimum prosthodontic rehabilitation.

- Early diagnosis and delay in hope that teeth will erupt should be discouraged. Early surgical exposure of teeth allowed normal root formation and eruption
- Treatment should be sequential with removal of deciduous teeth & surgical exposure of unerupted permanent teeth at time corresponding to their normal time of eruption. Concurrent continuous orthodontics is carried out. This regime maximizes the growth potential of alveolar bone, facilitates functional occlusion may obviate the need of orthognathic surgery
- Prosthodontic objectives are to address the underdeveloped maxillae alongwith increasing occlusal vertical dimension, establish functional occlusion, improved function, appearance and speech.
- Overlay prosthesis provided a optimum ,non-invasive prosthodontic solution for the situation discussed along with interdisciplinary care approach.

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