

A Dento – Surgical Management of a Cleidocranial Dysplasia Case. A New Approach for Better Oral Health Quality of Life Part II to: (A 9-year-old Saudi Boy with Cleidocranial Dysplasia)

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Abstract Background: Cleidocranial dysplasia is a rare congenital defect of autosomal dominant heritable disease with prevalence of 1 per 1,000,000 individuals. It is primarily affects bones which undergo intramembranous ossification [1,2,3,4]. **Case presentation:** A 9 years old Saudi male with CCD attended Pediatric Dental Clinics in Dammam Medical Complex. Chief complaint was delay eruption of permanent teeth and multiple caries. Based on clinical examination, radiographic investigations including CBCT & with coordination between Pediatric Dentistry, maxillofacial surgery and Orthodontic departments, a treatment plan has been concluded taking into consideration the wellbeing and the oral health related life style of the patient by addressing a new approach to preserve all the primary teeth, surgically removing all supernumerary teeth, applying removal or fixed modified Gropers' appliance to improve speech, esthetic & function than observing the eruption of the permanent teeth to intervene orthodontically only when needed. **Conclusion:** Team work involving multi-disciplinary departments helped to select a preeminent & new dental management approach for CCD patients including all aspects needed, Dental rehabilitation, Surgical & Orthodontic treatment to provide better oral health quality of life for the young patient.

Keywords: *cliedocranial dysplasia, dammam approach, oral health, quality of life, dento-surgical management*

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1. Introduction

Cleidocranial dysplasia (CCD) is a rare congenital defect of autosomal dominant heritable disease with prevalence of 1 per 1,000,000 individuals, equally affects both males and females. It is primarily affects bones which undergo intramembranous ossification [1,2,3,4]. It is also known as Cleidocranial dysostosis, osteodentin dysplasia, mutational dysostosis & Marie and Sainton disease, this skeletal disorder is first described in 1898 [5]. It is characterized by delayed closure of the cranial sutures, hypoplastic or

aplastic clavicles, cone shaped thorax, short stature with brachycephalic skull and bossing of the parietal and frontal bones and the presence of multiple dental abnormalities such as supernumerary teeth, retained primary teeth, delayed eruption of permanent teeth and enamel hypoplasia [6,7]. The skeletal relationship of the jaws tends to be in Class III position due to presence of a hypoplastic maxilla and decreased vertical growth due to poor development of the alveolar bone [8]. Besides defects in the skull and clavicles, CCD patients may suffer from other bony anomalies such as scoliosis, knock-knee and flat feet [9]. In addition, hearing loss and upper respiratory tracts infections are not uncommon in patients with CCD [9].

The definite cause of CCD is unknown, many studies have been conducted to determine mutations of the runt – related transcription factor 2 gene (RUNX2), located on chromosome 6p21 [10,11]. RUNX2 controls the differentiation of precursor cells into osteoblasts and is essential for intramembranous bone formation, which is associated with delayed ossification of the skull, clavicles, maxilla and teeth [6]. The gene is also required for mesenchymal condensation, osteoblast differentiation from mesenchymal stem cells, chondrocyte hypertrophy and vascular invasion in the developing skeleton [12,13]. This case report describes the clinical manifestations of CCD and treatment plan of 9.4 years old Saudi male patient presenting with mutation of RUNX2 gene.

when he is 11 years old and has been controlled by medication.

2.2. Clinical Features

The extra-oral examination showed frontal bossing, hypertelorism, depressed nasal bridge, convex profile, competent lip, his shoulder could be brought closer together and short stature (Figure 1). His medical history revealed delayed closure of the anterior fontanelle, absence of calvarian bone and partial occipital.

2. Case Presentation

2.1. Chief Complaint and Medical History

A 9 years old Saudi male attends Pediatric Dental Clinics in Dammam Medical Complex with his mother. The main concerns were about delay in the eruption of permanent teeth and multiple carious teeth. The patient has been diagnosed with Cleidocranial dysplasia since born. Patient developed recurrent episodes of convulsions



Figure 1. Cleidocranial Dysplasia

Tooth no.	Dental Problem	Tooth no.	Dental Problem
55	Occluso distal restoration with recurrent caries	75	Deep MODBL Caries
54	MOD Caries	74	Deep ODBL Caries
53	Labial incipient caries	73	Labial , lingual & distal caries
52	Remaining root	71	Mesial caries No mobility
51	Remaining root	81	Mesial caries No mobility
61	Remaining root	83	Labial caries
62	Remaining root	84	Deep ODBL Caries
63	Mesial caries	85	Deep MODBL Caries
64	OD deep caries		
65	OD deep caries		



Figure 2. Pre-Operative Photographs

Intra-oral findings showed normal gingival texture with melanin pigmentation, shallow alveolar bone and decreased vertical growth due to poor alveolar bone development, poor oral hygiene, multiple caries, hypoplastic teeth, retained primary teeth, delayed eruption of permanent teeth 31, 41, 36, 46, 16, 26, 32, 42, 11, 21, 22, 12, mesial step molar relationship and CL I canine relationship, overjet and overbite are not applicable due to upper anterior remaining roots.

3. Radiographic Findings

A panoramic radiograph revealed unerupted permanent teeth and supernumerary teeth in maxilla and mandible jaws, Dental age is 5 years old much delayed to the chronological age (9.4 years old) as eruption but as root formation dental age is 7 years old which is also delayed than chronological age. Bitewings radiographs and periapical radiographs were made to check proximal caries and any pathology related to roots and/or furcation area since the teeth are extensively decayed.

Cone beam CT scan (CBCT) were decided to correctly account and localize supernumerary teeth, it revealed in:

- The upper right posterior teeth: one permanent 1st, 2nd molar and three premolars.
- The upper right anterior teeth: one permanent canine, lateral incisor and two permanent central incisors.
- The upper left posterior teeth: one permanent 1st, 2nd molar and three premolars.
- The upper left anterior teeth: one permanent canine, lateral incisor and two permanent central incisors.
- The lower right anterior teeth: two permanent canines, two permanent lateral incisors one of them is lacerated and two permanent central incisors.
- The lower right posterior teeth: one permanent 1st, 2nd molar and two 2nd premolars and two 1st premolars.
- The lower left anterior teeth: two permanent canines, two permanent lateral incisors one of them is lacerated and two permanent central incisors.
- The lower left posterior teeth: one permanent 1st, 2nd molar and two 2nd premolars and two 1st premolars.

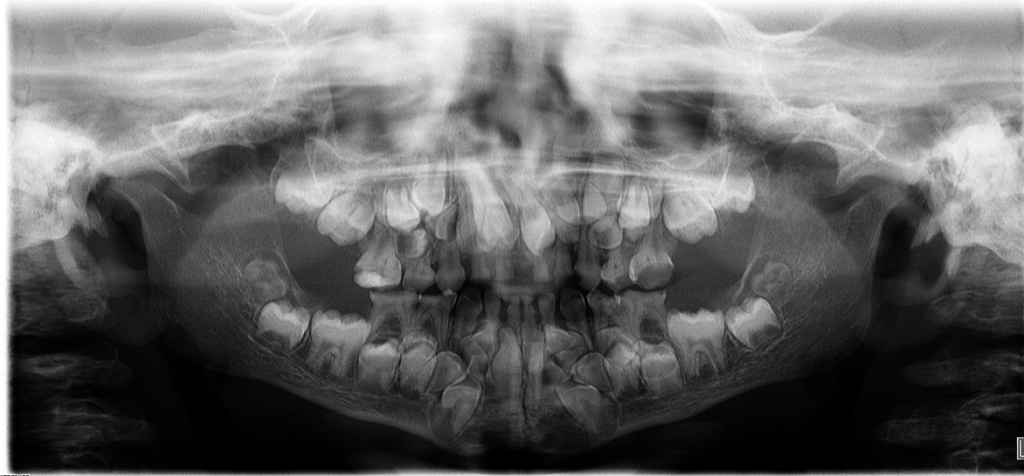


Figure 3. Panoramic Radiographs

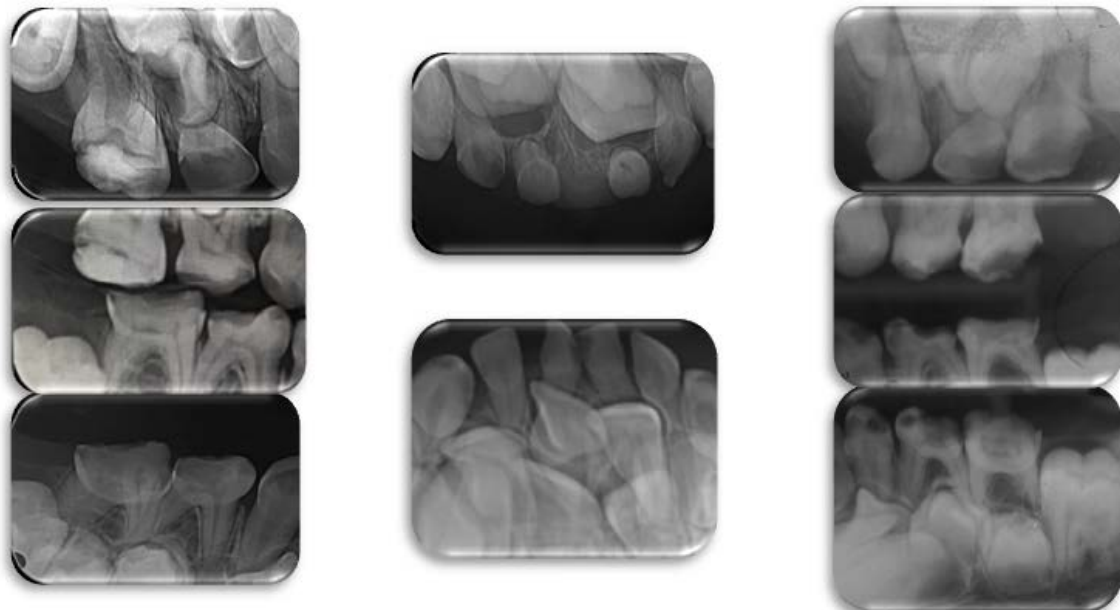


Figure 4. Pre- Operative Radiographs

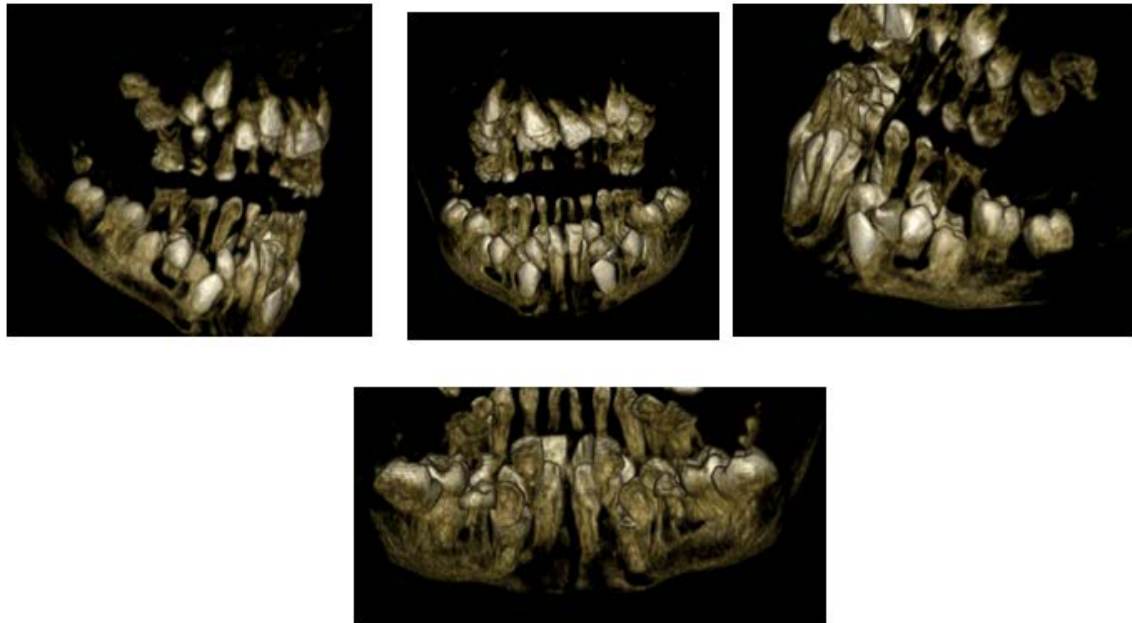


Figure 5. Pre- Operative CBCT

4. Treatment Plan

After taking the medical history of the patient, consultation was requested & cleared from his physician to exclude any contraindications for regular dental treatment settings or treatment under general anesthesia.

Treatment was planned & called (**Dammam approach**) as three steps:

The first step: dental management of all primary teeth & as the patient had definitely positive behavior (According to Frankle scale of behavior) all dental procedures were done in multiple dental visits & according to the American Association of Pediatric Dentistry (AAPD) Guidelines which involved restoring the function of the occlusion, improving the vertical distance, and to preserve all suitable teeth for orthodontic anchorage to tract anterior permanent teeth if needed. Follow up plan was tailored to improve oral hygiene of the patient, regular fluoride varnish application, diet control and dental home care routine.

According to Orthodontic consultation, extraction of the upper and lower primary anterior teeth was advised to accelerate the eruption of unerupted permanent anterior teeth.

The second step:

- surgical removal of supernumerary teeth in all quadrants were decided according to CBCT findings and interpretation to:
 - Count & locate the supernumerary teeth.
 - Selecting the well-formed teeth to be preserved
 - Access surgically without endangering any neighboring teeth germs, primary teeth roots & vital structures was the decisive factor in the decision making to preserve patient safety.
 - Give chance for spontaneous eruption of the developing permanent teeth with minimal orthodontic intervention when needed.
- Applying Modified gropers 'appliance to improve speech, function & esthetic of the patient.

The third step: Regular long term follows up:

- To give chance for spontaneous eruption of the developing permanent teeth without orthodontic intervention.
- To intervene orthodontically if needed by surgical exposure and/or traction.
- Orthodontic alignment of teeth after eruption if needed

There are different management approaches of patients with Cleidocranial dysplasia:

5. Dental Management

First step was done under regular dental setting involved complete dental rehabilitation:

Pulp therapy and stainless-steel crown. All upper anterior remaining roots and lower central incisors were extracted. The patient has been seen after nine months from dental treatment completion because the patients' caregiver was not compliant to dental appointments, his upper and lower permanent anterior teeth were still not erupted, occlusal radiographs were taken for the upper and lower anterior areas, it showed some progressive eruption movement of the labially placed lower central permanent incisors compared to the first occlusal radiographs.

The patient caregivers were firmly instructed to be more compliant with their son dental appointments, especially in his case to have best management and results.

The Second step: Patient was prepared for surgical removal of supernumerary teeth under general anesthesia as planned. The updated past medical history revealed that the patient had history of recurrent convulsions in the last three months and controlled by medication consequently consultation was made and cleared by Pediatric Neurologist.

In OR under GA, the patient was nasotreachally intubated. IV prophylaxis antibiotic was given.

In the lower jaw, full thickness envelop buccal mucoperiosteal flap is raised with mesial and distal

relaxing incisions from tooth #73 to 83, impacted teeth are exposed buccally with surgical bone removal and the following teeth were extracted, labial supernumerary of #31,41,32,42. Then from the lingual side an envelope full thickness lingual mucoperiosteal flap was raised from #74 to 84, the impacted teeth were exposed with surgical bone removal and extraction of supernumerary teeth of #45, 44, 34, 35, 83.

In the upper jaw, full thickness envelope buccal mucoperiosteal flap with distal and mesial incision was done from #54 to 64, palatal flap was raised as well. The impacted teeth were exposed with surgical bone removal, the following supernumerary teeth were extracted #11, 21, 52.

All the surgical wounds were closed with 4/0 vicryl, simple interrupted sutures with horizontal matrix. As shown in [Figure 4](#).

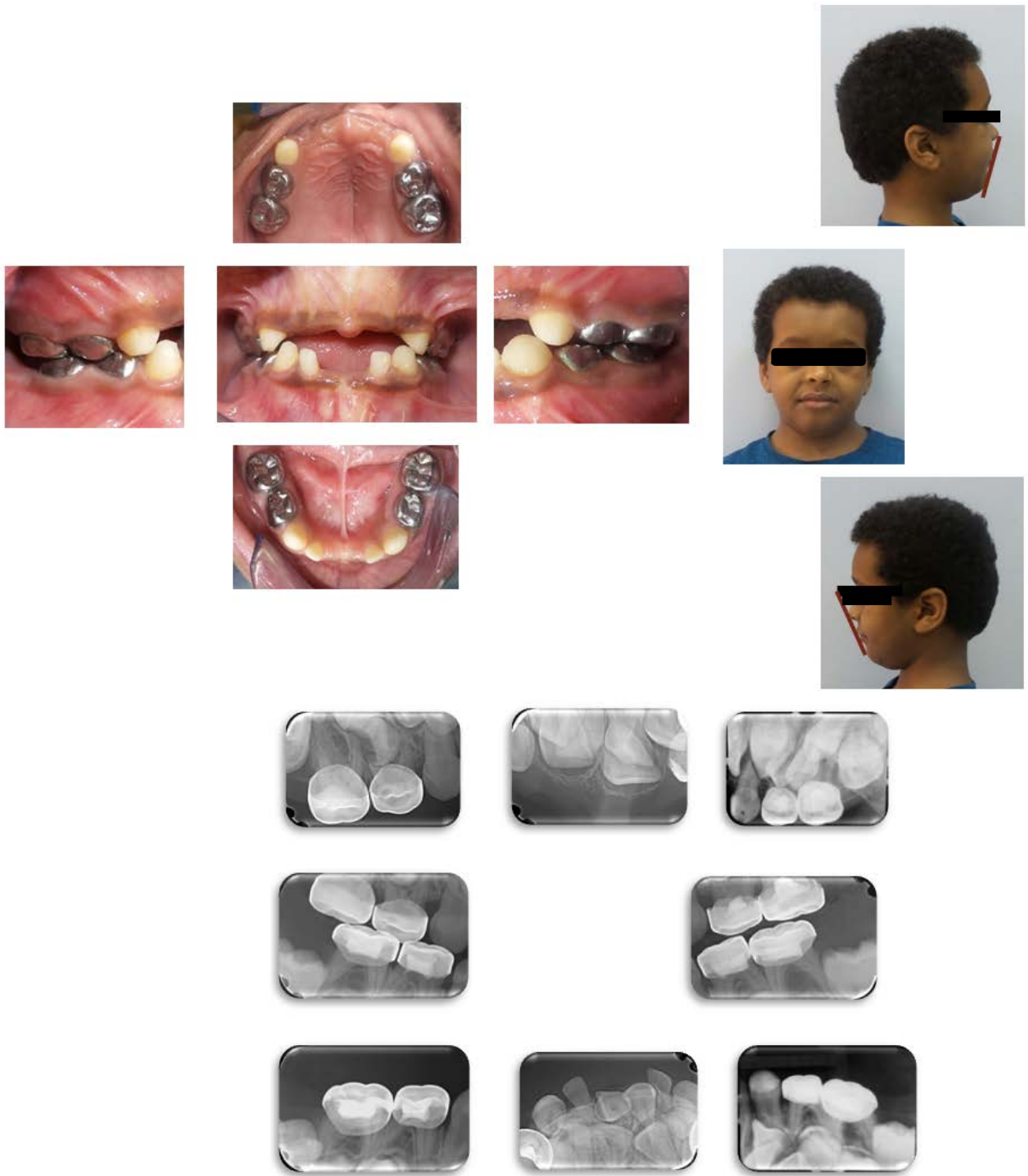


Figure 6. Pre-Operative Photographs & Radiographs

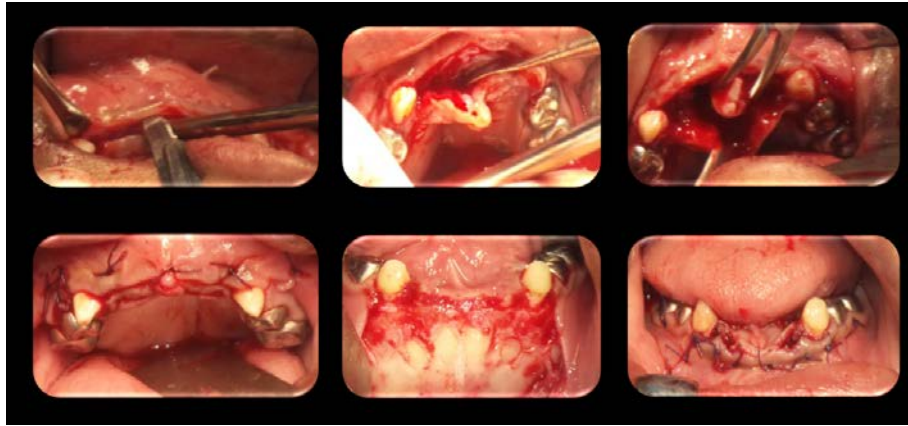


Figure 7. Some photographs are taken during the operation showing raising flap in the upper, extraction of palatal supernumerary tooth, Upper & lower simple interrupted sutures and lower labial supernumerary teeth



Figure 8. Post operation Cone Beam



Figure 9. Extracted Supernumerary teeth



Figure 10. Modified Gropers' Appliance

During the operation, we discovered some extra impacted supernumerary teeth, most of them were deciduous teeth at early stage of development and were superimposed on each other hence it was impossible to be seen in the previous CBCT. Some of impacted supernumerary teeth could not be removed as decided before the operation because their location may endanger the vital structure, permanent teeth follicles or roots of deciduous teeth. So, we decided to leave them at the moment and take another CBCT after the operation to be certain about the exact number and location.

The patient was seen after 10 days from the operation, soft tissue healed properly and there were no operation complications associated. Orthodontic Bands were selected for upper and lower E's then alginate impressions were made for the upper and lower jaws to construct modified Gropers' appliances with the aim to improve speech, esthetic and function.

The new CBCT revealed 5 extra supernumerary teeth, 4 in maxilla (2 in the right premolars area, 2 in the left premolars area) 1 in the mandible look as deciduous canine in the left side. So, the total number of the supernumerary teeth of this patient are 19 not 14 as previously thought.

As during the operation, we extracted 9 impacted supernumerary teeth in the mandible & 3 in the maxilla. Accordingly, there are 7 teeth left, need to be removed later (4 in the upper jaw and 3 in the lower).

Modified Gropers' appliance were adjusted and tried intraorally before cementation. Patient was comfortable and happy about the end result. The appliances were cemented with glass ionomer. The appliance can be used as fixed/removable without cementation depending on the oral hygiene of the patient [Figure 10](#).

Actually, the patient is on a follow up monthly basis to monitor permanent teeth eruption, oral hygiene and replace/adjust the appliance when needed according to the patient growth and needs.

6. Discussion

Cleidocranial dysplasia is a definite clinical phenotype arising from deregulation of intramembranous and

endochondral ossification due to a mutation in *Cbfa1* (*RUNX2*) which is located on the short arm of chromosome 6. Hypermobility of the shoulders, clavicles absence or short clavicles, incomplete cranial sutures closure, and supernumerary teeth are seen to be consistent features of Cleidocranial dysplasia [2-20]. In our case, the foremost features, like the absence of clavicles, broad skull sutures, partial occipital bone were present, in addition there was numerous impacted & supernumerary teeth with the consequences of delayed permanent teeth eruption.

The treatment objective in our case was to consider the oral health related quality of life of the young patient (**OHRQoL**) [23] in our decision making. Our dental management & treatment plan was focused on the improvement & maintenance of patient psychological, social & functional needs.

The literatures of CDD Dental management revealed 4 different approaches as shown in [Table 2](#), started with **Toronto-Melbourne approach**: The first phase starts in very early age from 5-6 years old which is not applicable in our case and has multiple phases of surgical interventions that may affect the patients' compliance and expose him to unnecessary frequent surgical exposure. Regarding **Belfast-Hamburg approach & Bronx approach**: both require some previously erupted permanent teeth in place for orthodontic anchorage to facilitate the eruption of the other impacted teeth which is also not applicable in our case because none of the permanent teeth were erupted yet. As well as, both approaches compromise patients' oral health quality of life and are more convenient for cases in older age or after growth termination. **Jerusalem approach**: was the nearest applicable approach to our case but we modified it according to (**OHRQoL**) basis. The main modifications are delaying orthodontic traction for (6-12 months) and observing the eruption of permanent teeth after surgical removal of supernumerary teeth, as the crowding will be relieved & allow for the permanent teeth to erupt, in addition; the root development of unerupted permanent teeth, as shown in the recent CBCT, have already reached 2/3 or more and that will help in accelerating their eruption. Modified Gropers' appliance construction has been decided in the upper and lower arches to improve the speech, function & esthetic of the

patient while waiting for permanent teeth eruption, this new approach called: (**Dammam approach**) according to our city name as shown in [Table 1](#).

In spite of all well planned dental management, these cases always carry challenges and dilemmas that may affect the treatment plan path, but the treating team patience, knows when to stop, how to evaluate and manage the estimated risks in every situation, is a corner stone in management success of such cases.

For example we could not remove all the impacted supernumerary teeth as a one shot & the fact that we

discovered new extra supernumerary teeth that were not been seen in the previous CBCT due to their superimposition, dictated us to change our decision and re-plan because the risk get over the benefit.

As a pediatric dentist, our role is to improve Oral Health Quality of patients 'life by providing both primary & comprehensive preventive treatment, as well as therapeutic needs [24], helping early detection of such cases, pointing the main problems and outlining the treatment plan in a multidisciplinary team work when needed.

Table 1. Dammam Approach

Management approach	Age	Procedure
Our new approach (Dammam Approach)	10-12 years	Phase 1: -Extraction of anterior primary teeth. - Extraction of all supernumerary teeth. -Construction of modified Gropers' appliance to improve esthetic . -Observe eruption of permanent teeth radiographically every 6mo-12mo. -Orthodontic traction when needed
	>13 years and older	Phase 2: - Extraction of posterior primary teeth. - Construction of Partial denture. -Observe eruption of permanent teeth radiographically every 6mo-12mo. - Orthodontic traction when needed

Table 2. Criedocranial dysplasia: Other management approaches Reference [21,22]

Management approach	Age	Procedure
Toronto-Melbourne Approach Reference: [14,15,16,17]	5-6 years	- Extraction of deciduous incisors. - Deciduous incisors are exposed and healing is allowed.
	6-7 years	- Orthodontic brackets are placed on permanent anterior teeth incisors. - Extraction of posterior deciduous teeth.
	9-10 years	- Permanent bicuspid are exposed. - Surgical removal of supernumerary teeth and healing allowed
	9-12 years	- Surgical exposure of permanent premolars - Brackets placed on canines and premolars.
Belfast- Hamburg Approach Reference: [18,19]	Single method Age: not specified	- All primary and supernumerary teeth are removed. - All impacted teeth are surgically exposed. - Surgical packs are placed to prevent healing of bone and soft tissue over teeth. - Healing by secondary intention. - Orthodontic attachments are placed. - Orthodontic appliances placed on fully erupted teeth. - Elastic thread is placed between brackets on unerupted teeth and the arch wires.
Jerusalem approach Reference: [15,16]	10-12 years	Phase 1: - Anterior primary teeth are extracted. - All supernumerary teeth are extracted. - Permanent incisors are exposed. - Orthodontic attachments are placed on permanent incisors. - Surgical flaps are closed completely.
	>13 years and older	Phase 2: - Posterior primary teeth are extracted - Unerupted permanent canines and premolars are exposed. - Orthodontic attachments are bonded. - Surgical flaps are closed completely.
Bronx approach Reference: [19]	Age: not specified	Phase 1: - All primary and supernumerary teeth are removed - Surgical flaps are closed. Phase 2: - Unerupted 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.

7. Conclusion

The facts behind achieving new alternative approach, are to improve oral health quality of life (OHQoL) for the young patient, by providing the basic psychological & social needs, like good esthetic, improve speech & function. Early diagnosis and knowledge of the clinical characteristics, family history and dental management in a team work coordination are the corner stone for having better management and prognosis in Cleidocranial dysplasia cases. The multidisciplinary team can involve Pediatric dentistry, Orthodontic and Maxillofacial departments, Pediatrics, Genetics and laboratory technician. Last but not least, patient compliance to the dental appointments is an important and necessary requirement.

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Ethical Consent

The protocol of the present case report was approved by the Ethical Committee at Eastern Province Directorate, Ministry of Health. The informed consent was agreed about by the patient caregiver (Father). All dealings performed in this case were in accordance with the ethical standards of the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Conflict of Interest

Authors declare no potential conflict of interest.

Abbreviations

- Cleidocranial Dysplasia: CCD
- General Anesthesia: GA
- Oral Health Quality of Life: OHRQoL
- Teeth Charting used: FDI double digit numbering system adopted by WHO

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