Surgical Management and Evaluation of the Craniofacial Growth and Morphology in Cleidocranial Dysplasia

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Abstract: Cleidocranial dysplasia (CCD, MIM 119600) is a rare autosomal dominant disorder affecting bone, cartilage, craniofacial growth, and tooth formation leading to supernumerary teeth. Few reports delineate the genotype–phenotype correlations related to the variations in craniofacial morphology and patterning of the dentition and the complexity of treating patient's malocclusion. Successful management of the craniofacial deformities in patients with CCD requires a multidisciplinary team of healthcare specialists. Approximately 70% of patients are due to point mutations in *RUNX2* and <20% due to copy number variations with the remainder unidentified. There is no literature to date, describing the orthognathic management of CCD patients with deletion in one of the *RUNX2* alleles. The purpose of this study was to evaluate the craniofacial morphology and dental patterning in a 14-year-old Caucasian female with CCD resulting from a novel microdeletion of *RUNX2* in 1 allele. The CCD patient with *RUNX2* haploinsufficiency due to microdeletion had decreased craniofacial bone and ankyloses in the permanent dentition. An altered extraction protocol of supernumerary teeth was followed in this patient. Craniofacial growth and morphologic analysis demonstrated atypical skull shape, persistent metopic suture, and decreased mandibular size.

Key Words: Cleidocranial dysplasia, cone beam computed tomography, superimposition, supernumerary teeth

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Cleidocranial dysplasia (CCD, MIM 119600) is a rare autosomal dominant disorder globally affecting 1 in 1,000,000 individuals and is caused by mutations in the transcription factor, RUNX2.[1–4] RUNX2 plays important roles in osteoblast differentiation, skeletal morphogenesis, chondrocyte proliferation, and differentiation and tooth formation.[2–5] Clinical significance of CCD includes short stature, clavicular dysplasia, wormian bones, patent fontanelles, hypertelorism, midfacial hypoplasia, short distal phalanges, scoliosis, genu valgus, and pes planus.[1–4] Craniofacial/dental manifestations in CCD include over-retained deciduous teeth, supernumerary teeth, and malocclusion. Clinical manifestations in patients with CCD include recurrent upper respiratory tract and ear infections, hearing loss, speech delays, dysphagia, dysarthria, and high risk of osteoporosis.[6–10]

RUNX2 point mutations cause approximately 70% of CCD patients, while <20% of patients result from copy number variation with the remainder of patients uncharacterized.[6] Recent reports demonstrate microdeletions (allelic heterogeneity) leading to abnormalities in the craniofacial complex solidifying the need for more studies evaluating the genotype–phenotype correlations of CCD patients.[7–14] Complex clinical challenges are associated with treating CCD due to adolescent growth patterns and the complexity of tissues involved.[15–20] Due to broad craniofacial and dental involvement, successful treatment requires a staged, multidisciplinary approach. Despite numerous advances in surgical and clinical techniques, challenges continuously arise related to delayed growth of the midface, failed tooth eruption, and numerous supernumerary teeth in CCD patients.

Treatment strategies for CCD involve multiple phases of care. Roberts et al[21] previously described established surgical approaches for managing the craniofacial and dental complex in patients with CCD, emphasizing in all patients the importance of early diagnosis and treatment. The most common surgical approaches for managing patients with CCD are the Toronto–Melbourne, Belfast–Hamburg, and the Jerusalem techniques.[21] The Toronto–Melbourne treatment strategy involves timed surgical extraction of all over-retained deciduous teeth and supernumerary with removal of overlying bone based on the succedaneous tooth's root development.[21] The Belfast–Hamburg uses 1 surgical treatment in which all supernumerary and deciduous teeth are extracted under general anesthesia.[21] The Jerusalem approach involves 2 distinct surgeries, first extraction of all anterior deciduous and supernumerary teeth; however, a 2nd surgery is undertaken around 13 years old where all remaining deciduous teeth and supernumerary teeth are removed.[21] At the time of the 2nd surgery, using the Jerusalem approach, all unerupted canines and premolar teeth are surgically exposed and bracketed for guided tooth movement into dental arch.[21] The Bronx approach is another technique that involves 3 surgeries comprising of removal of supernumerary and deciduous teeth, fabrication of esthetic removal partial dental prosthesis, and LeFort I osteotomy.[21] In this

TABLE 1. Cephalometric Landmarks

Landmarks	Description	Definition
A	A-point	Most concave aspect of the anterior maxilla
B	B-point	Most concave aspect of the anterior border of the mandibular symphysis
ANS	Anterior nasal spine	Anterior point of the maxillary bone
PNS	Posterior nasal spine	Posterior point of the maxillary bone
Ba	Basion	Anterior aspect of the foramen magnum
Me	Menton	Most inferior point of the mandibular symphysis
Pg	Pogonion	Most anterior point of the mandible
Go	Gonion	Most posterior inferior position of the angle of mandible
OP	Occlusal plane	Sella-Nasion to occlusal plane, angle formed
S	Sella	Medial aspect of sella turcica
N	Nasion	Most anterior position of frontonasal suture
SNA	Maxillary position relative to cranial base	Sella-Nasion to A point, angle formed
SNB	Mandibular position relative to cranial base	Sella-Nasion to B point, angle formed
ANB	Maxilla relationship relative to mandible	A point to B point, angle formed
MP	Mandibular plane	Plane from menton to gonion
FMA	Mandibular plane angle	Frankfort Mandibular plane, angle formed
LFH	Lower face height	ANS to menton, linear measurement
E-plane	Ricketts' E-line	Line between the nasal tip and soft-tissue chin point
U1	Upper incisor	Maxillary incisor position
L1	Lower incisor	Mandibular incisor position

study, we evaluate the craniofacial growth and morphology of a CCD patient with a rare *RUNX2* microdeletion undergoing active surgical and orthodontic care over a 5-year period of adolescent growth.

MATERIALS AND METHODS

Clinical and Radiographic Evaluations

The protocol for this study was approved by the institutional review board and the University of Alabama at Birmingham (UAB). Comprehensive health histories were gathered from conferring clinicians. Clinical examinations were accessed periodically following initial diagnosis and treatment plan by UAB Dental Genetics Clinical Team. Quarterly to biannual clinical and radiographic examinations were obtained using Nikon D70 6.1 MP Digital SLR Camera and cone beam computed tomography (CBCT). Superimposition analysis was performed using Invivo 5.2.3 (San Jose, CA) software of CBCT lateral cephalometric films of proband preoperatively and postoperatively. A list of common cephalometric landmarks and their definitions is shown in Table 1.

Clinical and Radiographic Findings

Medical history revealed bilateral hearing deficiencies and short stature. Preoperative craniofacial examination revealed disproportionate skull morphology, brachycephalic profile, parietal and frontal bossing, hypertelorism, patent frontal suture (Fig. 1), and joint hypermobility (ie, shoulders, not shown). The patient's oral examination revealed she was in mixed dentition with 11 overretained deciduous teeth (A, B, H, J, K, L, M, N, Q, R, and S), an anterior crossbite (9) and 6 unerupted permanent teeth (6, 7, 18, 19, 30, and 31) (Figs. 1 and 3). Radiographic examination further revealed open frontal suture, multiple wormian bones, hypoplastic hyoid bone, under-developed midface, abnormal dentoalveolar bone height with small mandibular ramus and body, and clustered supernumerary teeth in lower right (28A, 28B, 29A, 29B), left (21A, 21B, 21C), and upper right (4A, 5A) quadrants (Fig. 2).

Treatment Objectives and Protocol

The treatment objectives were to obtain esthetic and functional dentofacial rehabilitation for this patient. Orthodontic and surgical interventions, as outlined below, was used to guide eruption of impacted teeth, removal of supernumerary teeth, correct the dental malocclusion, and increase lower facial height. The comprehensive orthodontic treatment included preparation of dental arches (placement of labial and lingual arches) to facilitate guided eruption of surgically exposed teeth, correction of the crossbite and increasing the vertical dimension of occlusion by eruption of posterior teeth. The surgical treatment used a modified Belfast–Hamburg approach and was carried out in 2 stages. Stage I, in the upper arch treatment included, but was not limited to, extraction of upper left primary cuspid (H), extraction of upper right supernumerary coronal to upper right cuspid 6, and expose of the upper right and left lateral

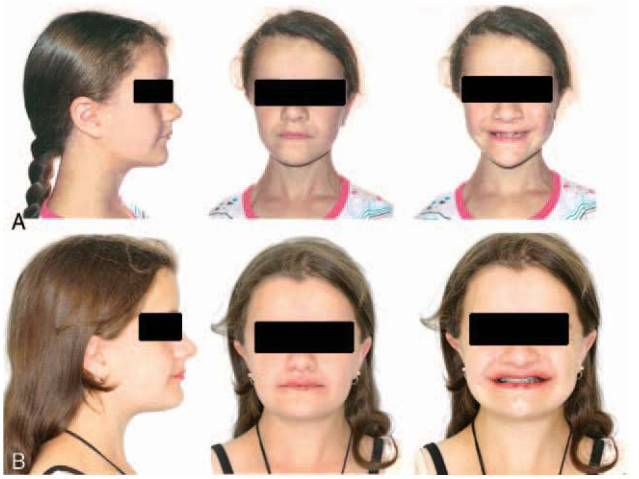

FIGURE 1. Facial photographs of the cleidocranial dysplasia patient: (A) pretreatment and (B) final. Initial photographs demonstrate morphologically brachycephalic, square-tapering profile, hypertelorism, low-set ears, and frontal bone depression. Final photographs demonstrate brachycephalic increased lateral growth and shorter lower facial third.

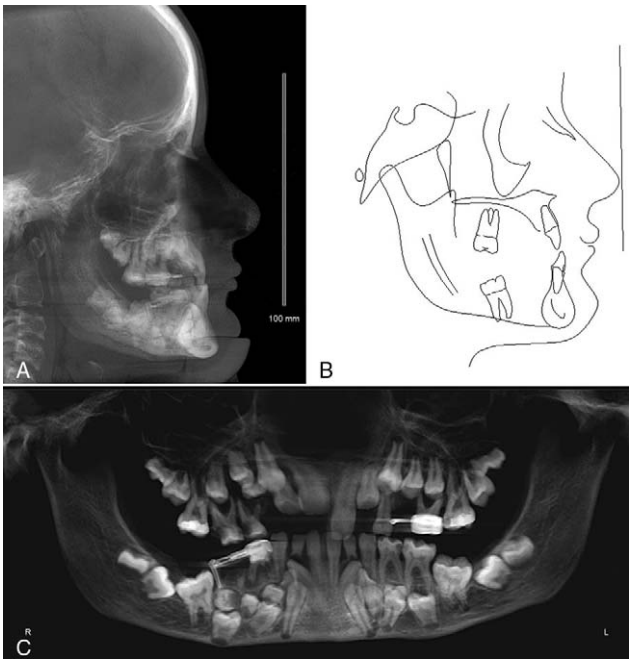

FIGURE 2. Initial radiographs derived from cone beam computed tomography. Lateral cephalometric radiograph (A) with tracing (B) and panoramic view (C) demonstrating multiple over-retained primary teeth, 7 supernumerary teeth predominantly in mandible and failed eruption of permanent incisors, premolars, and molars.

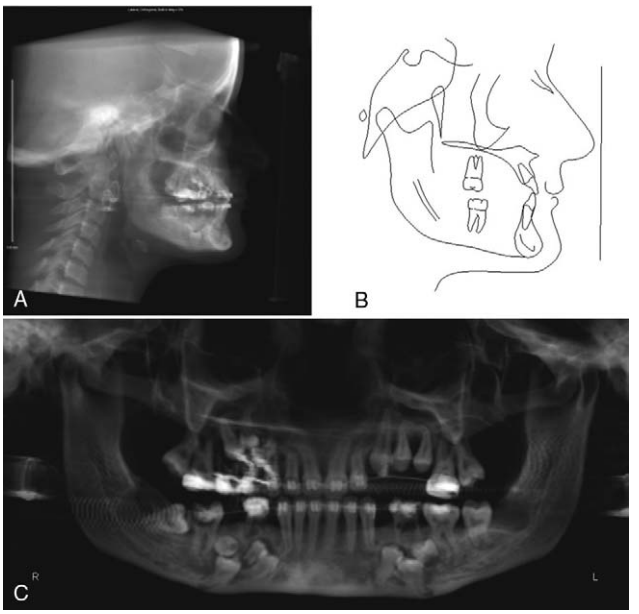

FIGURE 4. Final radiographs derived from cone beam computed tomography. (A) Lateral cephalometric; (B) lateral cephalometric tracing; and (C) panoramic view demonstrating remaining localized clustered supernumerary teeth in body of mandible, malpositioned right 2nd molar and failed eruption of multiple maxillary teeth.

incisors and cuspids (6, 7, 10, 11). In the mandible, extraction of primary teeth and supernumerary teeth above lower canine, as well as exposure of all mandibular anterior (22, 23, 26, 27) and permanent first molars (19 and 30) were planned for Stage I. Stage II involved extraction of remaining supernumerary teeth in the maxilla and mandible and exposure of permanent premolars and 2nd molars. All surgical treatment was performed under general anesthesia.

RESULTS

In this 5-year evaluation of a rare CCD patient with a total *RUNX2* deletion on 1 allele, multiple supernumerary teeth, and over-retained primary teeth were surgically removed, anterior permanent teeth were guided into the dental arches and the anterior crossbite was corrected. However, there was unsuccessful forced eruption of posterior teeth to establish adequate vertical dimension of occlusion (Figs. 3 and 4). A modified Belfast–Hamburg approach was used because of the inadequate dentoalveolar bone and midfacial hypoplasia (Figs. 2C and 4C). After the initial surgery, a decision was made not to extract the posterior mandibular supernumerary teeth due to poor bone density and height in the mandible. Due to financial limitations, patient refused any further surgical interventions, including bone augmentations procedures. Furthermore, a decision was made not to prolong the orthodontic treatment of this patient due to the high caries risk, significantly compromising the dentition (Figs. 3 and 4). A partial prosthesis would be incorporated into a revised treatment plan to establish posterior occlusion.

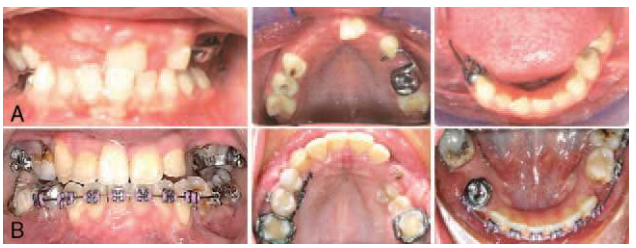

FIGURE 3. Intraoral photographs. (A) Initial frontal, maxillary, and mandibular photographs; (B) final frontal, maxillary, and mandibular photographs. Initial photographs demonstrate anterior crossbite, failed eruption of multiple maxillary teeth, failed exfoliation of multiple deciduous teeth. Final photographs demonstrate eruption of anterior teeth with minimal overbite and overjet, uprighting of lower molar and increased dental caries.

Because of the improvement in the anterior segments of the dentition, the upper and lower lip placement relative to the esthetic (E) line improved. The pre- and post-treatment (−4.9 to 8.0 mm and −5.8 to −8.2 mm, respectively; Table 2) values showed vast improvement the lower soft tissue profile gaining improvement in labial contour, whereas the soft tissue convexity showed no change.

Management of patients with CCD and other craniofacial, skeletal, and dental anomalies also requires cephalometric evaluation for determining optimal treatment planning and future recommendations. Furthermore, when surgery is under consideration a comprehensive assessment of the patient's growth potential is essential. In our CCD patient, cephalometric analysis revealed an increased in the ANB angle (from −4.1° to −2.0°, Table 2) decreasing the Class III relationship. The patient's facial axis was also evaluated using the angular relationship between 2 planes basion-nasion (Ba-Na) and foramen rotudum-gnathion (Pt-Gn). In normal Caucasian females, this value is 90°; however, in our CCD patient, the value was higher (100.2° and 96.4°, pre- and post-treatment, respectively) demonstrating a more anterior or protrusive growing chin. The mandibular plane angle (MPA) relative to SN (on average 32°) indicates whether there is an increase/excess or decrease/deficiency in vertical growth. In our study, the pre- and post-treatment MPA values (14.3° and 6.8°, respectively) illustrated significant deficiency in vertical growth and an overall hypodivergent growth. Furthermore, the Frankfort mandibular plane angle (FMA) provides an additional view of patients' vertical growth. The

TABLE 2. Cephalometric Analysis

	Initial	Dev Norm	5-Year Follow-Up	Dev Norm	Std Dev	Norm
Skeletal A-P						
SNA, °	87.6	1.6	92.0	2.8	3.5	82.0
SNB, °	91.6	3.2	94.0	3.8	3.4	80.9
ANB, °	−4.1	1.5	−2.0	−2.4	1.5	1.6
Skeletal vertical						
Facial axis (Ba-Na-Pt-Gn), °	100.2	2.9	96.4	1.8	3.5	90.0
Occlusal plane to SN, °	−8.4	−9.1	−6.5	−8.4	2.5	14.4
FMA (MP-FH), °	11.5	−2.8	4.1	−4.4	4.5	24.1
UFH (Na-ANS), %	40.6	−1.1	45.2	−0.2	5.0	46.0
LFH (ANS-Me || FH), %	59.4	1.1	54.8	−0.2	5.0	54.0
Dental						
Interincisal angle (U1-L1), °	155.2	4.2	137.0	1.2	6.0	130.0
U1-SN, °	102.9	0.0	124.2	3.9	5.5	102.8
Soft tissue						
Lower lip to E-plane, mm	−5.8	−1.9	−8.2	−3.1	2.0	−2.0
Upper lip to E-plane, mm	−4.9	0.4	−8.0	−1.0	2.0	−6.0
Soft tissue convexity, °	133.9	0.2	130.7	−0.4	4.0	132.4

A-P, anterior–posterior; FMA, Frankfort mandibular plane angle.

FMA angular measurements pre- and post-treatment in our study (11.5° to 4.1°, respectively) were significantly lower than norms (22.0° to 28.0°). The MPA changes, on average of 1° every 3 years, are well documented.[22–26] Based on normative values for our CCD patient relative MPA value should be approximate 17° at end of treatment (age 19); however, our patient's values are nearly 13° away from the norms demonstrating a significantly reduced to no vertical growth. In addition, the occlusal plane (OP) angle measured from OP to SN from pre- and post-treatment analysis (−8.40° to −6.5°, receptively) revealed a flat OP and lack of vertical growth despite the improvement in angular measurements. The posterior, upper, and lower facial height percentages (S-Go || FH, 86.2–92.9; Na-ANS, 40.60–45.20; ANS-Me || FH, 59.40–54.80) demonstrated counterclockwise movement of mandible with minimal changes over the 5-year period. Overall, the vertical indicators were all consistently depicting a skeletal vertical growth discrepancy, moderate brachycephaly, and hypodivergent mandible.

Cleidocranial dysplasia proband's facial morphology was evaluated clinically by CBCT on pre- and post-treatment photographs and CBCT films. Superimposition analysis obtained using extrapolated 2-dimensional cephalograms taken pretreatment and 5-year post-treatment. Cephalograms superimposed on the cranial base, maxilla, and mandible. Growth analysis revealed growth of the posterior cranial base and an anterior and downward displacement of the maxilla but only an anterior displacement of the mandible relative to the cranial base (SN) (Fig. 5). Although there was some mandibular growth (at condyles), the significant growth of the posterior cranial base resulted in no overall growth in the vertical axis (Fig. 5).

Facial morphology was assessed using pre- and post-treatment clinical photographs (Fig. 1) and CBCT films (Figs. 6 and 7). Facial form 5-year post-treatment demonstrates brachyfacial morphology with increased growth in the lateral dimension. The patient's total facial height (N-Gn) demonstrated mild to no change in vertical dimension from pre- and post-treatment (94.5 and 94.1 mm, respectively; Table 3); however, facial width (Zy-Zy) (109.5 and 111.9 mm, respectively) demonstrates mild growth in the lateral dimension. Cone beam computed tomography evaluation of the calvaria revealed multiple wormian bones and a persistent metopic suture consistent with frontal bossing and depressed forehead. Analysis of the metopic suture demonstrated approximately 56% reduction (16–8.9 mm) in suture opening; however, it failed close during this 5-year study.

DISCUSSION

Craniofacial growth and morphogenesis are complex processes involving genetic, epigenetic, and environmental factors contributing to the development and differentiation of both mineralized and soft tissues. During normal development, assessments of growth can be accurately obtained by evaluating stable craniofacial landmarks: the cranial base, maxilla, and mandible.[23–26] Dental malocclusions in CCD patients are often due to inadequate arch length, failure of tooth eruption, over-retained primary teeth, and multiple supernumerary teeth. These patients have delayed craniofacial growth, prolonged treatment phases beyond teenage, and early adulthood.[23–26]

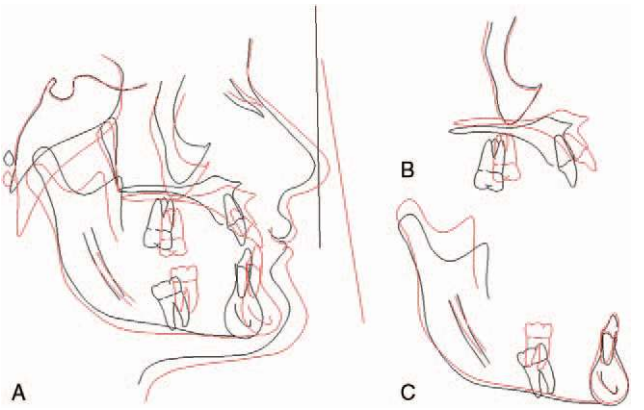

FIGURE 5. Superimposition analysis. (A) Cranial base superimposition demonstrates no vertical growth and minimal growth in the AP dimension; (B) maxillary superimposition reveals incisor and molar position improvements with minimal to no growth in the A-P dimension; and (C) mandible superimposition depicts vertical upright of molar and minimal growth posteriorly of the mandible. Pretreatment (black) and post-treatment (red). A-P, anterior–posterior.

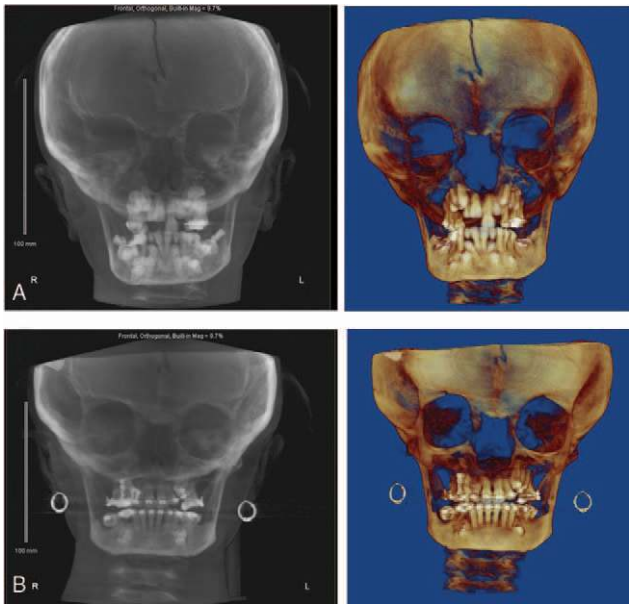

FIGURE 6. Cone beam computed tomography frontal view. (A) Pretreatment reveals persistent metopic suture, parietal and frontal bossing, mandibular cant with multiple supernumerary teeth; (B) post-treatment images demonstrate decreased metopic suture length, increased lateral cranial growth, and localized supernumerary remaining in body of mandible.

Management of the oral, dental, and craniofacial complex in CCD is cumbersome and requires a multidisciplinary team approach to include but not limited to surgeons, pediatricians, otolaryngologists, orthodontists, pediatric dentists, and speech therapists.

In this study, we used a modification of the Belfast–Hamburg approach due to the inadequate dentoalveolar bone and midfacial hypoplasia (Figs. 2C and 4C). Our goal was to use the remaining primary molar teeth (Fig. 2C) to establish and maintain the vertical dimension while surgically guiding impacted teeth (Fig. 4C). To date, few studies report the management, outcome, and craniofacial growth in patients with CCD. Evaluating growth and morphogenesis of patients with CCD undergoing active orthodontic and surgical management provides important insight into treatment progress, strategies, and outcomes, which are vital to patient care. Hwang et al[27] demonstrated successful clinical outcome following surgical correction of frontal bossing, hypertelorism, low nasal bride, midfacial hypoplasia, and mandibular prognathism using forehead

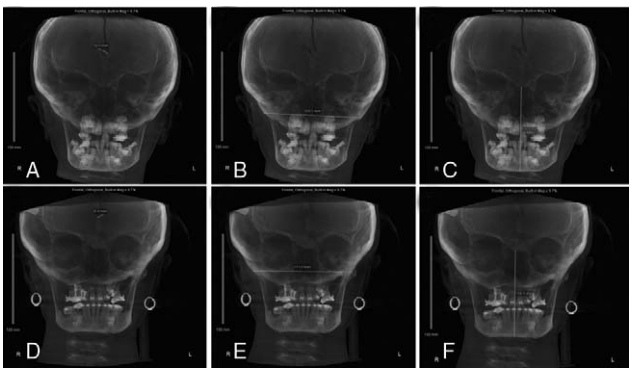

FIGURE 7. Cone beam computed tomography frontal view. Initial and final anthropometric measurements of the metopic suture (A, D), transverse (B, E), and facial height (C, F).

TABLE 3. Cleidocranial Dysplasia Cone Beam Computed Tomography Facial Proportions

	Initial	Final	Norm
Total facial height (N-Gn), mm	94.5	94.1	108 ± 6
Facial width (Zy-Zy), mm	109.5	111.9	130 ± 6
Metopic suture length, mm	16.0	8.9	0 (fused)

plasty, epicanthoplasty, augmentation rhinoplasty, malar and paranasal augmentation, and reduction genioplasty, respectively. In the report by Hwang et al,[27] although the classical skeletal features of CCD were corrected, the author did not report the management of the malocclusion and impacted and supernumerary teeth.

Çimen et al[28] used a modified Bronx approach in an 18-year-old male with CCD. Correction of the skeletal and dental discrepancies was accomplished in 2 surgeries, first extractions of all remaining primary and surgical guided eruption of impacted teeth followed by bimaxillary orthognathic surgery for the correction of the anterior open bite and class III malocclusion.[28] In our study, we followed a similar approach; however, we only extracted the deciduous teeth in the anterior mandible and maxilla to allow for surgical guidance impacted incisors and canine teeth and maintenance of vertical dimensions (Figs. 2C and 4C). Decision to leave remaining supernumerary teeth was due to proximity to mandibular canal and encroachment of inferior border of mandible (Fig. 4C). Subsequent maxillofacial surgery was not elected due to patient desires.

Jensen et al[29] reported the craniofacial morphology in 35 patients with multigenerational adults with CCD surgical or orthodontic treatment. Assessments of average facials' diagrams and films in CCD revealed increased diameter of the calvaria with consistent frontal bossing.[29] In our study, the upper facial morphology was consistent with the reported findings; however, the frontal suture remained open and significant lateral calvarial displacement was observed. Jensen[29] also demonstrated greatest changes in the cranial base; however, these findings contrast the results in our study. They noted the displacements along the foramen magnum, sella, and clivus; however, in our study, no noted changes were observed.[29] These inconsistencies in cephalometric findings may be associated with variations in anthropometric points; however, the existing paradigm of the cranial base being regarded as stable landmark remains due to minimal variations during growth and development.

Ishii et al[30] evaluated craniofacial morphology in 14 pediatric and adult Caucasians diagnosed with CCD who had no history of orthognathic surgery. Lateral cephalometric radiographs revealed that the pediatric patients had normal mandibular morphology and shape; however, the adults presented with mandibular prognathism and smaller lower facial height compared with the pediatric patients.[30] These authors proposed that the increased horizontal growth of the mandible in adults compared with children with CCD is possibly due to lack of vertical facial growth associated with failure of tooth eruption. In our study, we performed superimposition analysis following 5-year active orthodontic treatment to assess the skeletal, dental, and soft tissue growth. In our study, we observed that the proband's growth was delayed both dentally and skeletally. There was some anterior–posterior growth and changes were observed in the transverse dimension; however, no overall vertical growth was evident at the end of the 5-year study. At time of treatment, the patient's chronological age did not reflect the anticipated dentoalveolar development. These findings delineate a critical window of treatment for this CCD patient. Furthermore, our study suggests that delaying surgical and orthodontic treatment of similar patients would not be recommended.

Madeira et al reported the surgical and orthognathic care of a 21-year-old female CCD patient.[17] They revealed favorable outcomes when addressing class III skeletal and dental malocclusion.[17] In our study, clinical care began when the patient was 14 years old. Growth analysis demonstrates mild mandibular prognathism due to hypodivergent mandible and lack of posterior dental support. We were unable to achieve our initial objective of increasing the lower facial height and a revised plan is to incorporate prosthesis for posterior support. Early treatment of CCD has been shown to be effective at minimizing unfavorable mandibular growth[16,20]; however, in severe patients, orthognathic surgery may be required.[17–19] On the other hand, our patient's straight profile did not indicate that orthognathic surgery was necessary.

Park et al[19] reported surgical and orthodontic management in 2 patients with CCD using methodology similar to the Bronx and Belfast–Hamburg approaches. In our study, we report a 14-year-old Caucasian girl with similar craniofacial and skeletal features to those reported by Park et al but a different dentoalveolar phenotype (Figs. 1 and 2).[19] Park et al[19] demonstrated the effectiveness of the Belfast–Hamburg approach in the presence of congenitally missing teeth in the mandible,[19] which was the justification of our approach in this patient.

This is the first report evaluating the craniofacial morphology and dental patterning of a CCD patient with total *RUNX2* deletions on 1 allele. In the literature, CCD sutural development in relationship to specific *RUNX2* mutations have not been well documented. Many patients with CCD present clinically with frontal and/or parietal bossing and forehead depressions often benefiting from surgery.[18] It is known that Sex determining region Y box (SOX9), a transcription factor important in bone formation and chondrogenesis interacts cooperatively with RUNX2.[31–34] Numerous reports have shown that SOX9 inhibits RUNX2 expression and that SOX9 down-regulation results in increase RUNX2. Various SOX9 inhibitors (such as IL-1 and TNFα) may play an important role in minimizing failed suture development if administered.[32–34] The metopic suture generally closes during first year of life in healthy patients.[18] At 14 years old our CCD proband demonstrated a large opening in the frontal bone with a persistent metopic suture still present 5 years later. Because of this delayed suture closure in conjunction with frontal bossing, the facial morphology in this patient appears increased in the lateral dimension over time.

Providing care for patients with CCD requires individualized long-term treatment plans with comprehensive clinical and radiographic evaluations by the specialized multidisciplinary healthcare team.

In this study, a novel *RUNX2* gene microdeletion encompassing entire *RUNX2* gene on 1 allele presented unique challenges with both surgically and orthodontically. Although there are several surgical-orthodontic regimens including Toronto–Melbourne, Jerusalem, Belfast–Hamburg, Modified Belfast–Hamburg, and Bronx approaches, they each contribute a similar goal in correction of malocclusion. The precise surgical-orthodontic approach should be individualized based on differences in the age of patient, timed serial extraction of presenting deciduous and supernumerary teeth, extended root formation of permanent teeth, bone covering of the underlying permanent and supernumerary teeth, feasibility of the simultaneous artificial eruption with orthodontic appliance, and comprehensive treatment of orthognathic surgery and dental implants.

The planning of dentofacial goals in CCD varies from one to another individual depending on the availability of medical/dental specialists, needs of the patient, age at diagnosis, social and economic aspects, and different CCD mutations. Cleidocranial dysplasia may present with phenotypic variability depending on clinical manifestations of the disorder and the ongoing mutations in the determinant gene. The surgical-orthodontic approach of CCD is relied on the retained and dystopic supernumerary and permanent teeth. Diagnosing and treating at an early age seems to obtain the better results of the dentofacial correction. It is important to discuss with patient that the treatment may be extensive and of long duration, which may be modified due to multiple factors such as finances, dental caries, or poor apical bony base. Individual surgical-orthodontic approach or technique is a challenge to apply for every circumstance of a CCD patient. Besides genetic counseling, the rationale of the treatment goal is to assist the unerupted permanent teeth and facial growth for acceptable esthetic and functional outcomes.

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