CASE REPORT

Dental team management for a patient with cleidocranial dysostosis

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Cleidocranial dysostosis is a rare autosomal condition that affects ossification. The dental abnormalities associated with it present a remarkable challenge in orthodontic treatment planning. Early diagnosis is extremely important to give the patient the best treatment options. Patients with cleidocranial dysostosis require a team approach with good communication and cooperation from the patient. Timing of the intervention is critical, and many surgeries might be required. The patient in this report was treated with a team effort that involved several dental specialties to achieve an optimal result. (Am J Orthod Dentofacial Orthop 2005;128:110-7)

Cleidocranial dysostosis is an autosomal dominant inherited condition; the classic feature is at least 1 clavicle partly or completely absent. The skull has a brachycephalic appearance, and the closure of the sutures and fontanelles is delayed compared with normal. The fontanelles exhibit secondary ossification centers forming wormian bones.[1,2] The skull can have parietal and frontal bossing.[2] The patient tends to be short in stature with a narrow chest and sloping shoulders. The jaw skeletal relationship tends to be Class III with a hypoplastic maxilla and a closing mandibular rotation. Vertical facial growth is decreased with poor vertical development of the alveolar bone.[2]

The dental features include lack of exfoliation or resorption of deciduous teeth, delayed eruption of permanent teeth, failure of teeth to erupt, multiple supernumerary teeth, and tendency for cyst formation of the unerupted teeth.[3] Often, it is advisable to uncover and bond the permanent teeth when extracting the deciduous teeth because extraction of the deciduous teeth does not consistently promote eruption of the permanent teeth.[4]

HISTORY

The patient, a white girl 10 years 10 months of age, was referred by her pediatric dentist for an initial orthodontic exam (Figs 1-3). She did not have a chief complaint. Her medical history was positive for a diagnosis of cleidocranial dysostosis. She was in good health with no medications, no known drug allergies, and no contraindications for dental treatment. Her temporomandibular joints were asymptomatic with normal range of motion and function. There was no history of trauma to the mouth, teeth, lips, or jaws. She was in the early mixed dentition, and complete orthodontic records were obtained.

MEDICAL DIAGNOSIS

The patient had been diagnosed with cleidocranial dysostosis with a history of multiple bone fractures. Most fractures involved her feet and upper extremities. Her clavicles were absent, and she could oppose her shoulders anteriorly in the midline. She had frontal bossing and a short stature. She also had left thoracic scoliosis that progressed enough to warrant treatment with a thoracolumbosacral orthosis brace, which was worn around the torso but did not extend to the head and neck region.

DENTAL DIAGNOSIS

Pretreatment facial photographs (Fig 1) showed a relatively symmetric mesocephalic frontal appearance with a straight profile and competent lips. She had a good chin-throat length with an acute nasolabial angle. The intraoral photographs and dental casts (Figs 2 and 3) showed a transitional Class III malocclusion with the permanent first molars in crossbite and maxillary and mandibular arch perimeter deficiencies.

The pretreatment lateral cephalometric radiograph (Fig 4, *A*) and cephalometric analysis showed a skeletal Class III relationship, deepbite tendency, and upright incisors. The posterioanterior cephalometric radiograph (Fig 4, *B*) showed no gross asymmetries. The panoramic radiograph and occlusal films all showed many

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Fig 1. Pretreatment facial photographs.

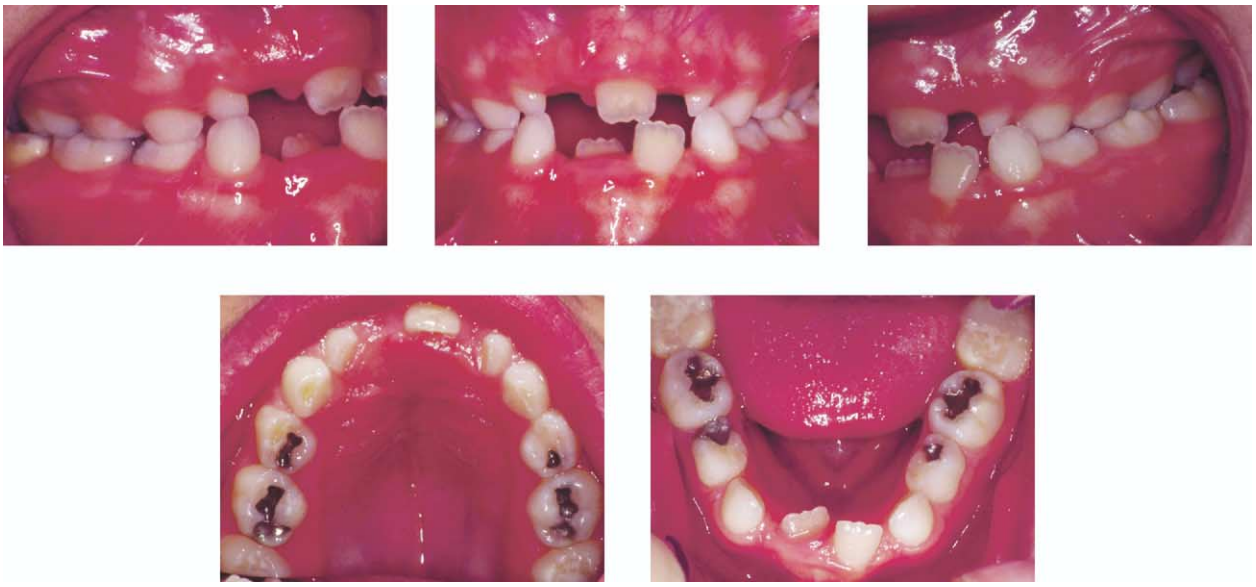

Fig 2. Pretreatment intraoral photographs.

supernumerary teeth and delayed eruption of the permanent teeth (Fig 5). No caries or other pathology was noted.

ETIOLOGY

This patient's malocclusion and many supernumerary teeth were due to the inherited characteristics of cleidocranial dysostosis. The etiology is an apparent spontaneous mutation. There was no history of facial trauma or digit sucking habits.

TREATMENT ALTERNATIVES

Treatment usually begins with the removal of deciduous teeth and supernumerary teeth to help the permanent teeth erupt. Several options are available for treating patients whose permanent teeth do not erupt on their own. Some are more costly and time intensive than others, and each patient's situation must be considered unique.

One option is to remove all permanent teeth and fabricate partial or full prostheses.[5] Another option is to surgically expose unerupted teeth to provide support for

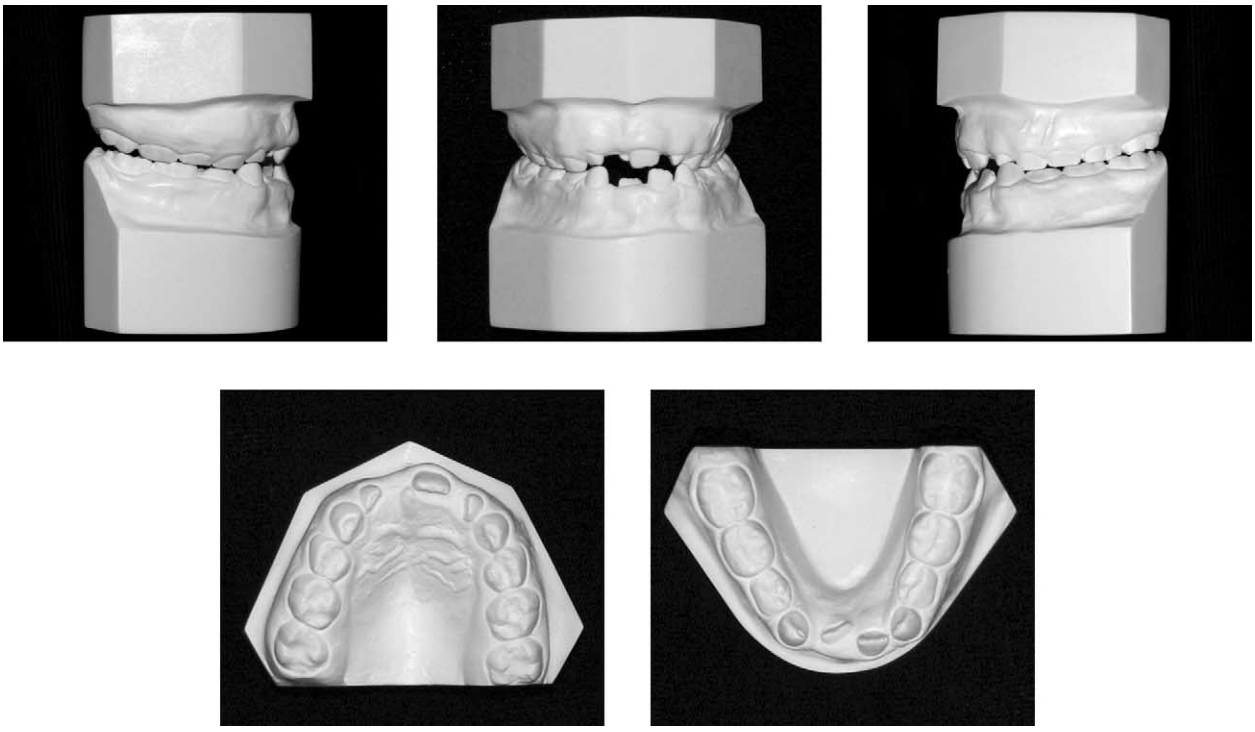

Fig 3. Pretreatment dental casts.

an overdenture.[6] The overdenture could also be supported by implants.[7] Impacted teeth could be translocated or relocated surgically.[2] Becker et al[8,9] presented an orthodontic and surgical method (the Jerusalem approach) that involves 2 stages of surgically uncovering and bonding the permanent teeth. The first stage is to guide the incisors into occlusion, and the second stage is to guide the remaining permanent teeth into occlusion. The use of implants in orthodontics as absolute anchorage also provides new opportunities to guide impacted teeth into occlusion.

TREATMENT OBJECTIVES

The treatment goals for this patient were to (1) correct the posterior crossbite; (2) surgically extract supernumerary teeth and remove anterior deciduous teeth; (3) uncover and bond the unerupted incisors and maxillary canines and guide them into occlusion; (4) monitor space needs and the Class III skeletal growth pattern; (5) extract the posterior deciduous teeth, uncover and bond the mandibular permanent canines and all premolars, and guide eruption into the arch when two-thirds of the root development was present for the permanent mandibular canines and all premolars; and (6) align and retain.

TREATMENT PROGRESS

The treatment plan proposed was very similar to what has been described by Becker et al[8,9] as the Jerusalem approach. The treatment objectives were explained to the patient and her parents, and informed consent was obtained. Phase I treatment consisted of a quad helix appliance for expansion and limited fixed appliances. Six maxillary and 4 mandibular supernumerary teeth were extracted by an oral surgeon, and the impacted incisors and maxillary canines were uncovered, bonded, and guided into the arch with traction. During the surgery, it was found that the maxillary permanent laterals were malformed; they were removed (Fig 6).

After the anterior teeth had erupted and been brought into alignment, the orthodontic appliances were removed, and the patient was placed in observation. Several incisors required gingivectomy by a periodontist to expose the entire clinical crowns. The remaining deciduous molars were extracted, and the mandibular permanent canines and all premolars were uncovered and allowed time for natural eruption.

At age 15, phase II treatment was initiated because the remaining permanent teeth had not erupted sponta-

Fig 4. A, Pretreatment lateral and **B,** posteroanterior cephalometric radiographs.

neously. Orthodontic appliances were placed on the anterior teeth and molars along with a transpalatal arch. Several unerupted premolars and mandibular canines were exposed and bonded. Because the patient had a dental and skeletal Class III relationship, it was decided to extract the mandibular second premolars and finish with Class III molar and Class I canine relationships. The mandibular second premolars were removed, instead of the first premolars, because their eruption and root development were significantly delayed, and they were adjacent to additional supernumerary teeth that had to be removed.

Because the maxillary lateral incisors were malformed and had to be removed, we developed a plan to replace them with implants and crowns. Movement of the maxillary canines was necessary to create the proper space and root divergence to accommodate implants. The risk in moving these teeth was discussed; the patient had dilacerations on some roots, especially on tooth #23 (FDI tooth number), and dilacerations have been associated with an increased risk of root resorption.[10]

The permanent first molars on the right were still in crossbite; this was corrected with crossbite elastics. The midline was corrected, and approximately 5.5 mm of space was created for the implants replacing teeth #12 and #22. Appliances were removed, and bonded wire retainers were placed on the maxillary central incisors and the 6 mandibular anterior teeth. A maxillary Hawley retainer was placed with pontics for teeth #12 and #22.

At age 17, the patient underwent surgery to remove her impacted third molars and place Brånemark Mark III 3.3 narrow body x 15-mm implants to replace teeth #12 and #22. Six months later, after osseointegration, the oral surgeon uncovered the implants. Transfer abutments were placed and confirmed with radiography to be fully seated. Impressions were taken, custom abutments fabricated, and porcelain crowns cemented by a prosthodontist. Almost 2 years after implant placement, tooth #21 became discolored and a radiograph showed that both teeth #11 and #21 had apical radiolucencies. The maxillary central incisors had necrotic pulps, and root canal treatment was performed by an endodontist.

TREATMENT RESULTS

Overall, the patient had an acceptable orthodontic result that should remain relatively stable (Figs 7-11).

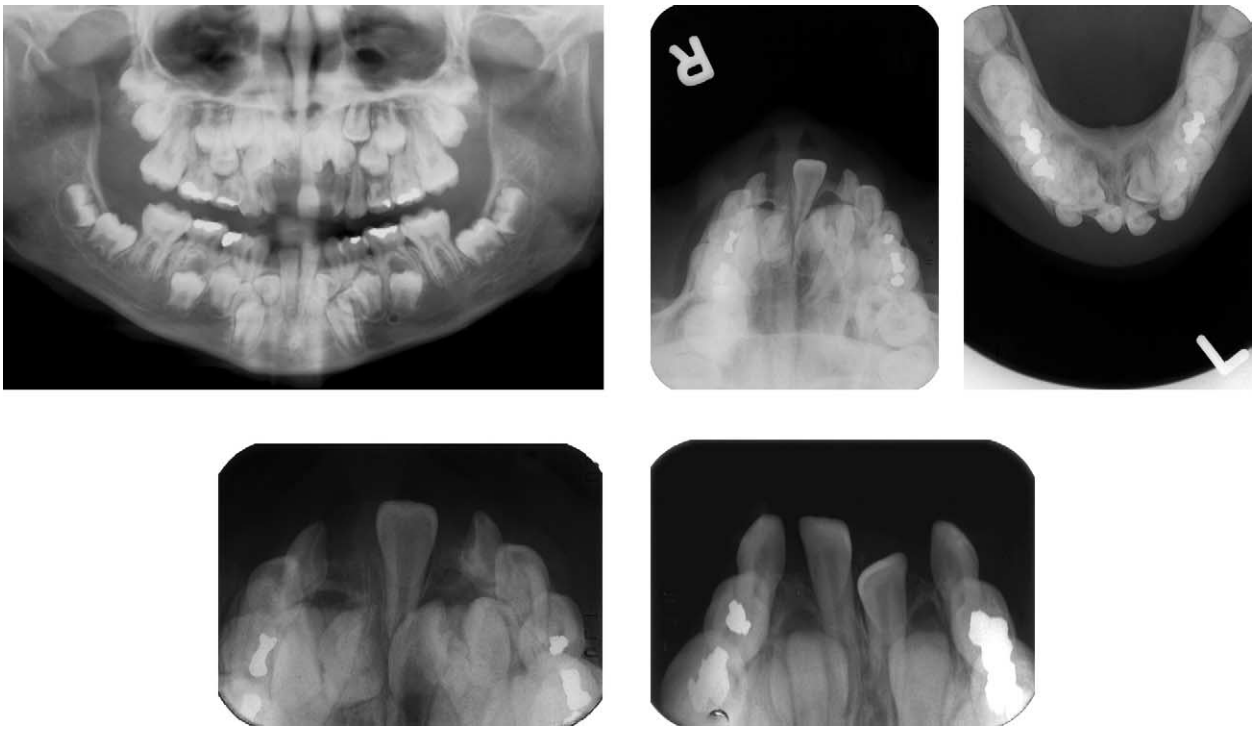

Fig 5. Diagnostic panoramic and occlusal radiographs.

Fig 6. Progress panoramic radiograph.

The overbite and overjet were idealized with coincident midlines. The implants and crown restorations for teeth #12 and #22 resulted in a pleasing outcome that will last her for many years. Because of the underlying skeletal Class III relationship and the removal of the mandibular second premolars, she finished with a Class III molar relationship bilaterally. The mandibular third molars, mesioangularly impacted because of insufficient posterior arch length, had to be extracted. However, the maxillary second molars finished in occlusion with the distal marginal ridges of the mandibular second molars, thus preventing any supraeruption over time.

The cephalometric superimpositions (Fig 12) showed that the mandible grew forward with little downward vector, in conjunction with minimal vertical facial development. The incisors were proclined; this improved the interincisal angle. The mandibular plane (to Frankfort) and SN to GoGn angles both decreased with growth. The profile changed as the nose and chin grew, resulting in decreased lip prominence.

DISCUSSION

A patient with cleidocranial dysostosis presents many treatment challenges to a dental team. The most challenging part of this patient's treatment was the unerupted permanent teeth, which were uncovered and guided into the arch with traction mechanics. This patient also had many supernumerary teeth that had to be removed. In addition, the permanent maxillary lateral incisors were dysmorphic, and a decision was made to remove them and replace them with implants after the patient reached skeletal maturity.

Surgical exposure of the unerupting permanent teeth with orthodontic guided eruption was the method of choice for this patient. This allowed her to keep her own

Fig 7. Posttreatment facial photographs.

Fig 8. Posttreatment intraoral photographs.

teeth and avoided the need for a prosthesis that would have to be maintained or replaced several times during her lifetime. The patient and family were pleased from both functional and esthetic standpoints. Other treatment modalities, including removable prostheses and surgical relocation, have disadvantages such as poor prognosis and results that can deteriorate over time.[2]

The orthodontic and surgical method was similar to the Jerusalem approach presented by Becker et al in 1997.[8,9] This method exposes the permanent teeth in 2 stages, allowing the patient to have the permanent incisors showing sooner, and the posterior permanent teeth when the time is appropriate. The 2-stage surgical interventions were planned for specific times that depended on the extent of root development. Similar to the Jerusalem method, the first intervention was initiated about age 10 to 12 years, when the anterior permanent teeth have two-thirds of their root development.[8,9] The anterior deciduous and supernumerary teeth were removed surgically. The permanent incisors and, in this patient, the maxillary canines, were exposed, and orthodontic attachments were bonded to the teeth with chains for traction. A closed-eruption technique was used to uncover the teeth.[11]

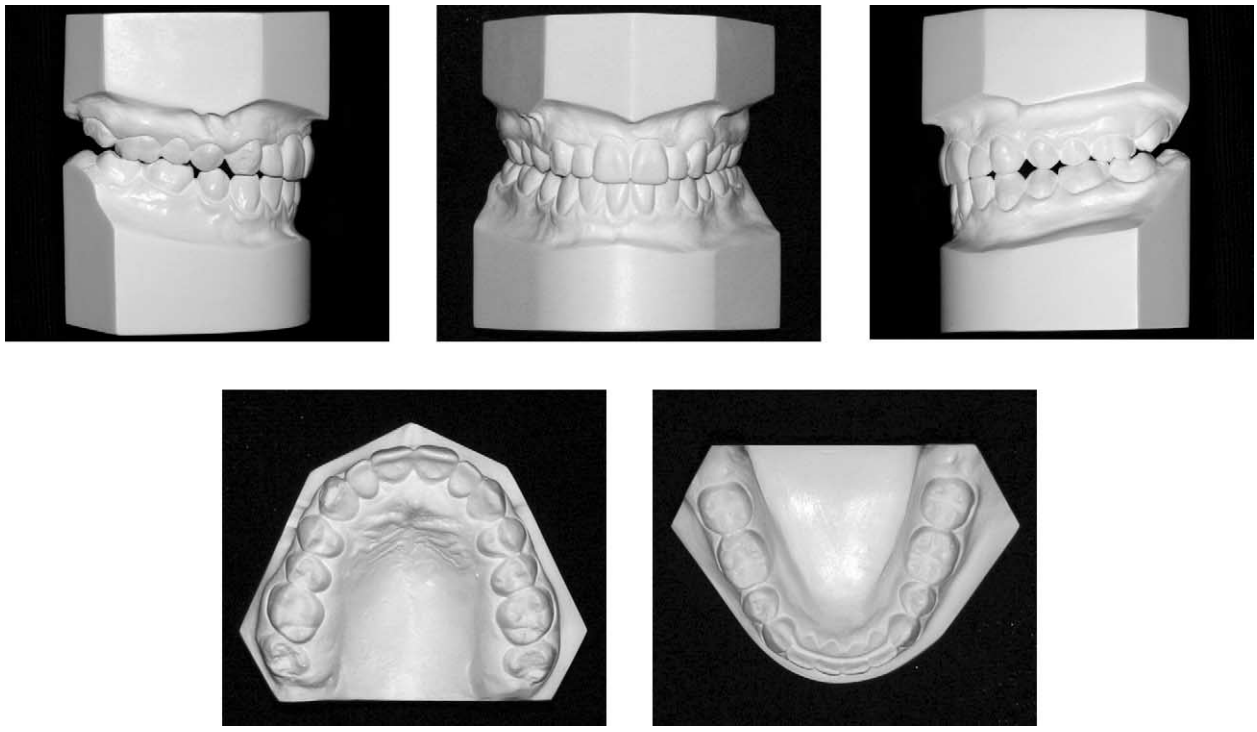

Fig 9. Posttreatment dental casts.

Fig 10. Posttreatment lateral cephalometric radiograph.

Fig 11. Posttreatment panoramic radiograph.

The second surgical intervention took place, again similar to the Jerusalem method, between 13 and 15 years of age, when the canines and premolars had two-thirds of their roots developed.[8,9] Bone was conserved during exposure of the canines and premolars, and attachments were bonded for orthodontic traction. The two-stage approach allowed the orthodontist to help the patient's self image by erupting the anterior teeth at the appropriate time and enhancing her smile.

The orthodontic portion of this treatment approach requires adequate anchorage, which was accomplished by banding the permanent first molars, placing brackets

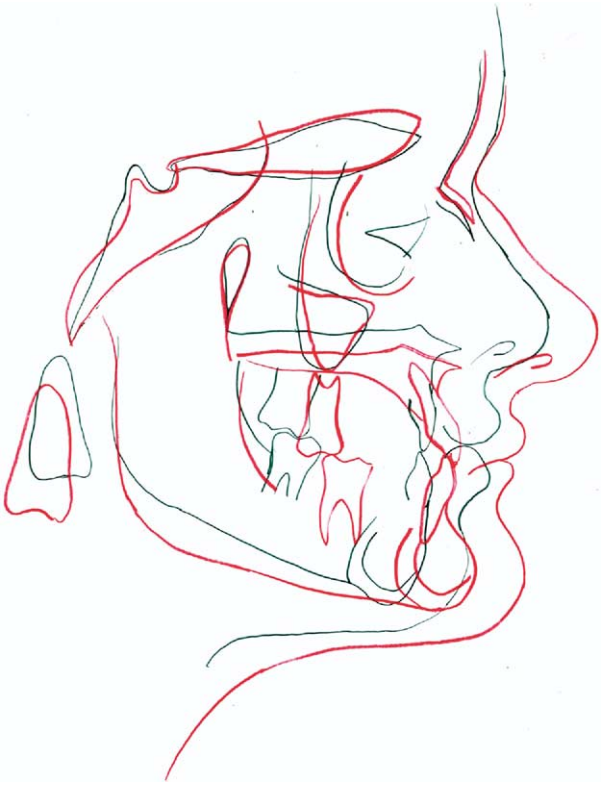

Fig 12. Lateral cephalometric superimposition.

on deciduous canines and molars, and placing a transpalatal arch. Light continuous forces were used to guide the impacted teeth into the arch and into alignment. In the second stage, the anterior teeth, permanent molars, and a transpalatal arch were used as anchorage. This patient was in treatment for approximately 8 years, so it was important to monitor and provide ongoing oral hygiene instructions to keep her dentition healthy.

The maxillary second molars had not yet fully erupted into occlusion at the patient's most recent visit, but we expect that the mesial cusps of the maxillary second molars will articulate with the distal of the mandibular second molars as they continue to erupt. If the mandibular third molars had not been impacted, it might have been more appropriate to maintain them to occlude with the maxillary second molars.

Because the incisors were proclined during treatment and the maxillary lateral incisors were missing, retention consisted of a maxillary Hawley with pontics for teeth #12 and #22 and a fixed lingual wire retainer on the mandibular anterior teeth. The patient was monitored for cessation of growth by taking serial lateral cephalometric radiographs when she believed she had stopped growing and then again 6 months later. The 2 radiographs were traced and superimposed to confirm the lack of further growth. She was then referred to the oral surgeon for placement of implants in the areas of #12 and #22, and removal of her third molars. After the implants were placed and the maxillary lateral incisor crowns cemented, active treatment was considered complete.

CONCLUSIONS

Treating patients with cleidocranial dysostosis requires a dedicated and knowledgeable team of dental specialists. Timing is critical. The orthodontic treatment can be lengthy and requires good cooperation from the patient for the best outcome.

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