ACP
AMERICAN COLLEGE OF PROSTHODONTISTS®

Interim Prosthetic Phase of Multidisciplinary Management of Cleidocranial Dysplasia: "The Bronx Approach"

Robert W. Berg, DMD,[1] Kenneth S. Kurtz, DDS, FACP,[1,2] Ikuya Watanabe, DDS, PhD,[3] & Anthony Lambrakos, DDS[1]

[1]Advanced Education Program in Prosthodontics, New York University College of Dentistry, New York, NY
[2]Division of Prosthodontics, Department of Dentistry, Montefiore Medical Center/Albert Einstein College of Medicine, Bronx, NY
[3]Department of Dental and Biomedical Materials Science, Nagasaki University Graduate School of Biomedical Science, Nagasaki, Japan

Keywords
Cleidocranial dysostosis; cleidocranial dysplasia; magnetic retention; cosmesis.

Correspondence
Kenneth S. Kurtz, DDS, FACP, Departments of Dentistry and Otolaryngology, Montefiore Medical Center/Albert Einstein College of Medicine, 3332 Rochambeau Avenue, Bronx, NY 10467. E-mail: ksk4@nyu.edu

Presented as a Table Clinic at the 2008 American College of Prosthodontists Annual Session by Dr. Lambrakos and as a lecture at the 2010 ACP Annual Session by Dr. Kurtz.

Accepted August 19, 2011

doi: 10.1111/j.1532-849X.2011.00786.x

Abstract

This case report presents treatment of two patients with the usual characteristics of Cleidocranial Dysostosis. A multidisciplinary approach using the disciplines of prosthodontics, orthodontics, and oral surgery was effected. Exfoliation of the patient's deciduous teeth and failure of permanent anterior tooth eruption led to emotional, social, and self-esteem issues in both patients. Due to the psychosocial issues confronting these two patients, esthetics was addressed prior to active intervention with orthodontics and after some surgical intervention. The use of two interim overdenture prostheses with magnetic retention is described.

Cleidocranial dysplasia (CCD) is an autosomal dominant skeletal dysplasia characterized by hypoplastic/aplastic clavicles, brachycephalic skull, midface hypoplasia, delayed closure of fontanelles, and moderately short stature. The estimated prevalence of CCD is one per million, but it is most likely under diagnosed because of the relative lack of medical complications in comparison to other skeletal dysplasias.[1-3] CCD is reported in all ethnic groups, and there is no sex predilection.[4,5] The chromosome related to the genetic expression of CCD is the short arm of 6p21, which is the locus for the RUNX2 gene. This is the "Master Gene" for the formation of bone and dental tissues. Transcription factor CBFA1 mutations are also associated with CCD.[6]

Dental manifestations are the major cause of morbidity. Most commonly reported findings include delayed or failed eruption of permanent teeth and existence in both jaws of multiple supernumerary teeth.[7] Other dental findings may include failure of deciduous tooth exfoliation, submucous cleft palate, hypoplastic maxilla, and malocclusion.[8] These dental findings typically necessitate multidisciplinary treatment that may span several years. Several combination treatment approaches have been reported.[9,10] Three distinct orthodontic/surgical approaches have been reported with great success including the Toronto–Melbourne approach,[11,12] the Belfast–Hamburg approach,[13,14] and the Jerusalem approach.[10,15]

The goal of any dental treatment should include the elimintion of the dental etiology of the associated morbidity, facilitating function, and improvement of the patient's appearance. The Toronto–Melbourne Approach,[11,12] a multiple surgical/orthodontic approach (Table 1), involves the removal of deciduous teeth, dependent upon root development of the permanent teeth, in multiple stages. The surgical procedures are the same, but at different time intervals. The first surgical procedure occurs around 5 to 6 years at which point the deciduous incisors are extracted. The deciduous posterior teeth remain until a later stage, usually around 9 to 10 years of age. The incisors are exposed at around the time the first molars spontaneously erupt, usually much later, packing is placed, and a period of healing is allowed prior to placement of orthodontic brackets. At around 9 to 12 years, a similar procedure is performed to extract remaining posterior deciduous teeth and to expose the premolars. This approach mentions no details of the orthodontic mechanics involved.

The Belfast–Hamburg approach,[13,14] (Table 2) is a single surgical approach that limits the number of surgeries to a single episode. All deciduous and supernumerary teeth are extracted,

Table 1 Multiple surgery approach (Toronto-Melbourne)

Series of surgical procedures Removal of deciduous teeth under general anesthesia (timing dependent on root development of permanent dentition) No description of orthodontic mechanics	
Stage 1	**Surgery**
5 to 6 years	Extractions/deciduous incisors
9 to 10 years	Extractions/deciduous posterior teeth
Stage 2	**Surgery/Orthodontics**
When first molars have been banded	Sugical exposure of permanent incisors
Following healing from previous surgical exposure	Brackets placed on incisors
9 to 12 years	Surgical exposure of permanent premolars/Supernumerary teeth removed
Following healing of previous surgical exposure	Brackets placed on premolars and canines

Table 2 Single surgery approach (Belfast-Hamburg)

Limit surgery to one episode Removal of all decidudous and supernumerary teeth and exposure of unerrupted teeth simultaneously (General anesthesia) Surgical packing placed and replaced to prevent bone healing and soft tissue closure over exposed teeth Brackets placed when healing has occurred and clean field for placement

Table 3 Two surgery approach (Jersusalem)

Two surgical interventions (timing dependent on root development of permanent dentition) Removal of deciduous teeth under general anesthesia (timing dependent on root development of permanent dentition)	
Intervention 1	**Surgery/Orthodontics**
Dental age 7 to 8	Extractions/anterior deciduous teeth and all supernumerary teeth Exposure of permanent incisors Brackets bonded immediately Surgical flaps closed
Intervention 2	**Surgery/Orthodontics**
Dental age 10 to 11	Extractions/remaining deciduous teeth Exposure of unerupted premolars and canines Brackets bonded immediately Surgical flaps closed

Table 4 Surgical-prosthetic approach (The Bronx)

One major surgical intervention (timing dependent on root development of permanent dentition) Removal of deciduous teeth under general anesthesia (timing dependent on root development of permanent dentition) Interim partial overdenture to maintain cosmesis	
	Surgery
Intervention 1	Extractions/deciduous teeth and all supernumerary teeth Surgical flaps closed
Intervention 2 (If necessary)	Exposure of unerupted teeth Brackets bonded immediately Surgical flaps closed
Intervention 3	Leforte I osteotomy-orthognathic surgery Placement of dental implants

and all unerupted permanent teeth are exposed simultaneously under general anesthesia. Surgical packing is placed, and healing occurs by secondary intention. The surgical packs are changed frequently until brackets are able to be bonded into place. Orthodontic appliances are placed on the few fully erupted teeth, and elastics are tied to the unerupted teeth to encourage eruption.

A third approach, the Jerusalem approach[10,16] (Table 3), involves two planned surgical interventions with their timing dependent on root development of the permanent teeth. The first planned procedure involves extracting all the anterior deciduous and all supernumerary teeth, exposing the permanent incisors and bonding orthodontic brackets immediately with full closure of the surgical flaps at the age of 10 to 12. The second surgical procedure takes place around age 13+ years. The remaining deciduous teeth are extracted, unerupted premolars and canines are exposed, orthodontic brackets bonded, and surgical flaps fully closed. These stages are carried out simultaneously in both jaws under general anesthesia. One of the goals of this approach is to immediately deal with the absence of anterior teeth by placing an orthodontic appliance to erupt the anterior teeth first.

The three above-mentioned surgical/orthodontic combined approaches do not address achieving some level of cosmesis for their patients during the long course of treatment. In the Jerusalem approach, initial efforts are concentrated towards bringing the anterior teeth into the mouth early, an attempt to provide a natural, age-appropriate appearance and improve the patient's self-image.[10,16] However, the duration of time to surgically expose and erupt the impacted anterior teeth is long. The dental age of these patients also lags 3 years behind their chronological age. This paper describes a fourth approach, an intervention that uses an interim overdenture prosthesis during the long course of treatment at Montefiore Medical Center; therefore, the authors refer to this treatment as the "Bronx Approach" (Table 4). Recognizing that this paper describes two successful treatment outcomes, we propose that further successful use of this treatment could ultimately lead to this approach being considered along with the other previously listed approaches for the treatment of CCD.

Figure 1 Initial extraoral and intraoral presentation.

Figure 2 Patient demonstrating absence of clavicles. Patient with and without prosthesis.

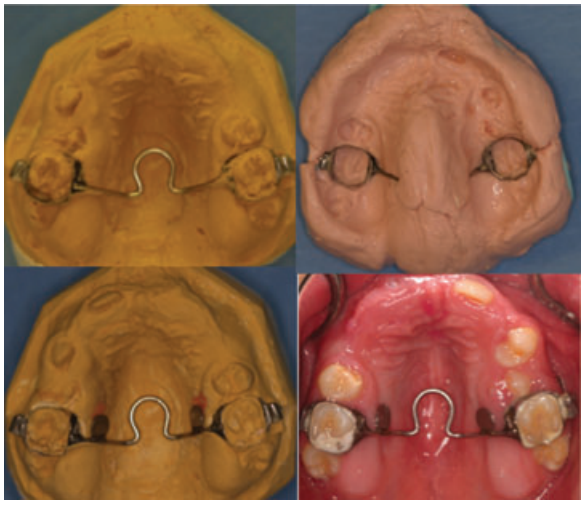

Figure 3 Counter-clockwise from top right: Intraoral pickup impression of molar bands; Initial cast; magnetic keepers laser welded to transpalatal arch wire; intraoral view of cemented appliance.

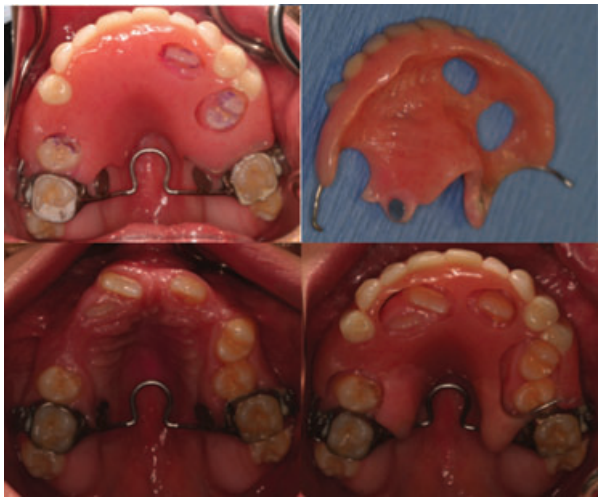

Figure 4 Clockwise from top left: Occlusal view overdenture without magnets attached; cameo surface overdenture with magnets attached, intraoral occlusal view with and without prosthesis.

The dental literature suggests that surgical/orthodontic treatment outcomes are favorable; however, the management of these patients may be difficult, because they may not appear in our offices at the times recommended for specific stages of treatment, and lack of coordinated care may modify the clinician's approach. Despite this challenge, the goals of treatment remain the same: preserving or restoring a proper functioning masticatory system and improvement of the patient's appearance. Increasing occlusal vertical dimension and establishing a functional occlusion will dramatically improve appearance, function, and speech of these patients. Restoring cosmesis will improve the patient's mental outlook and psychosocial image.[17] Lengthy, invasive, and sometimes unpredictable treatment involves a complex issue: informed consent for the adolescent. As a minor, the child must rely upon an adult parent or guardian to grant permission for different healthcare practitioners to provide elective treatment, and the prosthodontist must discern whether it is the parents' or the child's request for "normalcy." Certainly, it is difficult to ethically ignore a syndromic child's request for cosmesis, yet costs of providing coordinated

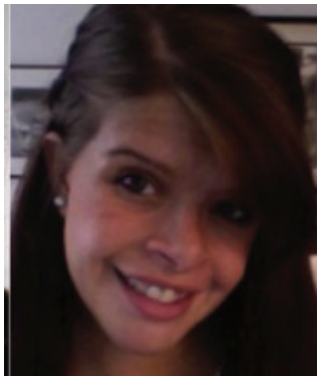

Figure 5 Patient after treatment.

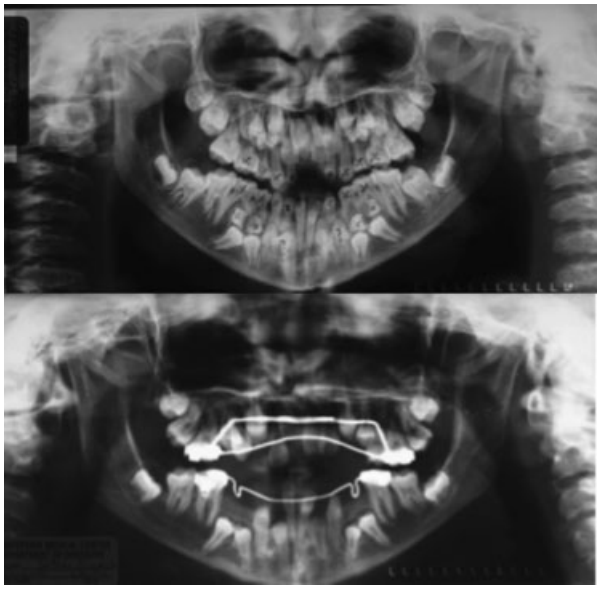

Figure 6 Initial radiographic appearance and orthodontic progress-guided tooth eruption each arch.

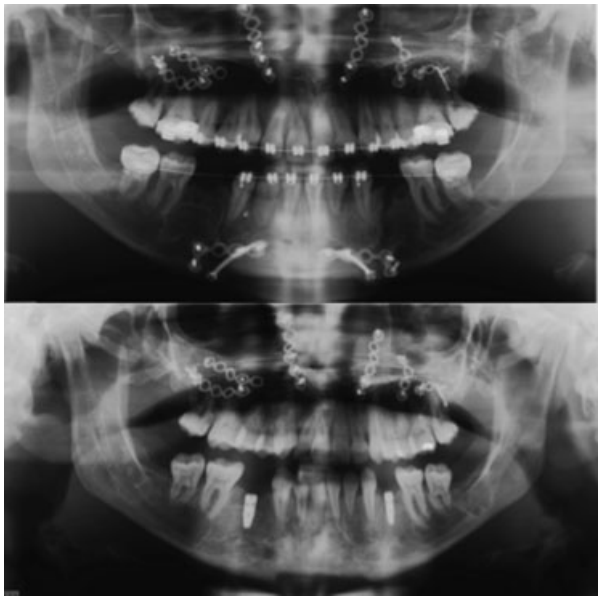

Figure 7 Panoramic radiographs post-orthagnathic surgery (Lefort I and genioplasty) (top) and post-endosseous implant placement (bottom).

multidisciplinary care are considerable, as are the time periods of active, staged treatment.

Clinical report

Two patients with the usual characteristics of CCD presented to the authors for treatment of their dental issues. The chief complaint of both patients were similar. The foremost issue both patients wanted to address was the lack of anterior teeth, causing them difficulties assimilating into their schools' social environment. In other words, they wanted to "fit in and be just like everyone else. I want normal teeth." Due to the psychosocial issues confronting these two patients, esthetics were addressed before intervention with orthodontic and surgical specialties. Both patients had a major surgical event under general anesthesia to remove all of the deciduous teeth and supernumerary teeth before them presenting at our clinic.

Due to the complex nature of these patient presentations and need for extensive oral surgical and orthodontic intervention, need for change in vertical dimension, and maxillofacial prosthodontic treatment, they were classified as type IV according to the Prosthodontic Diagnostic Index classification system for partially edentulous patients.

Typically in this patient population the molar segments of the dentition tend to erupt on time. This allows the prosthodontist to use orthodontic appliances to gain a mechanical advantage to retain a maxillary prosthesis. In both of these treatments, orthodontic bands were placed on the erupted maxillary first molars and connected with a transpalatal arch wire. Stainless steel magnetic keepers were laser welded onto the transpalatal arch appliance (TPA) bilaterally in different planes. Both patients exhibited hypoplastic maxillae, and this 3D deficiency readily permits placement of artificial teeth anterior to the maxillary alveolus, helping achieve superior esthetics and lip support. Artificial tooth arrangements were assessed intraorally and were approved by the patients. In our opinion, actively involving the teenage patient in the treatment process helps them to regain self-esteem. After acceptance of the artificial tooth arrangement, the overdentures were processed and finished in heat-polymerized polymethylmethacrylate (PMMA) resin. Closed field Neodymium-Iron (Nd-Fe-B) magnets (7.2 N attractive force, Hicorex, Hitachi Metals, Tokyo, Japan) 4 mm in diameter were attached to the prostheses intraorally with autopolymerizing PMMA resin (Jet Repair Acrylic, Lang Dental Manufacturing Inc., Wheeling, IL). Palatal windows perforating the denture base were created in the areas of the erupting permanent teeth so as to not interfere with the teeth as they erupted. The resulting prosthetic display of artificial teeth provided adequate cosmesis for each patient. Upon eruption of the anterior permanent dentition and sufficient posterior anchorage, the patients initiated conventional orthodontic treatment with full multibanded edgewise appliances.

Figures 1 to 11 detail the course of treatment for both patients described in this clinical report.

Discussion

Regardless of the approach to treatment of patients with CCD, surgery and orthodontics are inevitable. The various approaches differ in the number and timing of surgeries and when teeth are bracketed for orthodontic movement. Basic fundamental surgical and orthodontic principles are consistent with all of the approaches. Surgical treatment in this patient population may involve complications relating to multiple oral surgical events for CCD patients (Table 5).

The major difference between the newly introduced approach and the previous approaches is the maintenance of cosmesis throughout treatment. Treatment of the two patients in this clinical report took place over many years following the initial

Figure 8 Patient extraoral and radiographic initial appearance.

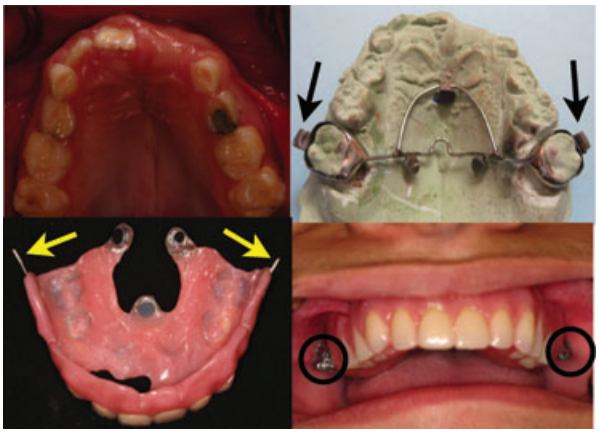

Figure 9 Top: Intraoral occlusal view, working cast with transpalatal arch appliance with magnetic keepers attached. Bottom: Intraoral view of buccal tubes engaged with wrought wire buccal clasps.

Figure 10 Cameo surface of prosthesis demonstrating the three different magnetic planes.

surgical intervention. They both presented at about 13 years old. The overriding concern for patients in the early adolescent and adolescent years is the need for cosmesis. As prosthodontists, we have the ability to provide this for the adolescent patient while also providing proper speech and function. Previous approaches, other than the Jerusalem approach, do not consider cosmesis in which they initially attempt to bring the anterior dentition into the oral cavity first.

One of the biggest challenges of prosthodontic treatment of these patients is retention of the maxillary prosthesis before and/or while patients are undergoing orthodontic therapy. This suggested technique offers multiple advantages. A TPA is almost ideal in this situation as the TPA does not restrict maxillary growth and will not interfere with the eruption of the permanent dentition. It will also maintain the position of the first molars, preventing mesial rotation and maintaining space in the arch.[18,19] First, the use of TPA creates an undercut for the wrought wire clasps of the overdenture to engage with to create indirect mechanical retention. Prefabricated orthodontic bands with buccal tubes attached can allow for direct mechanical engagement of the wire attached to the denture base. Orthodontic headgear is used in this same manner. Upon engagement of the buccal tubes on the bands, the overdenture's horizontal path of placement mechanically seats the prosthesis in the horizontal plane, and then the magnets can stabilize the base in the other two planes. Second, the TPA allows for the attachment of magnetic keepers that will actively attract the magnets attached to the intaglio surface of the overdenture. The prostheses designed in both patient treatments are similar in that they both are retained by magnets and wire mechanical retention. In the first treatment, the wrought wire buccal clasps provide lateral, resilient retention, while in the second patient treatment, they provide lateral, rigid retention; however, they differ in the arrangement and number of magnets. By placing magnets in different planes, it is possible to magnify the force and retention of the prosthesis. With the use of two magnets, a magnetic fulcrum exists. By adding an additional third magnet it eliminates the magnetic fulcrum.[20]

The challenges with the proposed approach include the frequent need for follow up and adjustment of the prosthesis. The prosthesis can interfere with the eruption of the permanent dentition if not adjusted and relieved frequently as the teeth erupt. Patient compliance is very important, as is parental understanding of this long process. Until a sufficient number of teeth have erupted and sufficient anchorage is present to bring the anterior

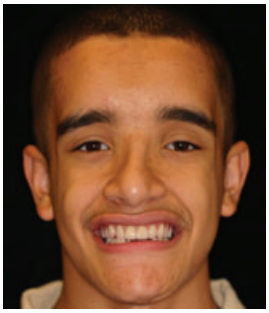

Figure 11 Patient smile with prosthesis.

Table 5 Complications associated with multiple surgical procedures in CCD patients

Tooth buds damaged by exposure trauma
Replacement resorption in crowns of impacted teeth
Surgical exposure of periodontal ligament/Ankylosis
Compromised bony support/Reduced periodontal integrity
Development of new supernumerary teeth (mid-teens)

teeth into the oral cavity, the prosthesis will remain in place and require modification. When the patients in this report reached this point in treatment, we were able to bond denture teeth to the traditional edgewise orthodontic appliances to maintain the cosmesis until the permanent dentition was aligned.

Regardless of patients' genetic conditions, self-esteem in the teenage years is very important to normal daily function. Syndromic patients are affected with issues related to the inability to assimilate within social environments due to their congenital differences. Prosthodontics is a discipline that enables these patients to have a sense of normality or likeness to their peers by providing them with a coping mechanism—a stable maxillary prosthesis. This can enable them to thrive socially. In the treatment of both patients, it was very gratifying to restore the confidence and esteem necessary for these adolescents to return to school and thrive. Historically, only a generation ago, the radical removal of all teeth for patients with CCD was routinely accomplished: "Generally the prognosis of orthodontic treatment is considered poor so that prosthodontics is the treatment of choice."[21] Aggressive treatment choices are required, and in patient treatment 1, 24 natural teeth were erupted (14-Max, 10-Min), a corrective Lefort I orthognathic surgery was accomplished, and in the mandibular premolar area, where teeth could not be erupted, single implants were placed bilaterally. There are currently two reports in the scientific literature, detailing the use of dental implants in patients with CCD.[15,22]

Certainly the eruption of the natural teeth, with their associated proprioceptive apparatus, allows the patient to experience a nearly natural dentate state. The single-tooth implants bilaterally placed in the mandible are a conservative effort to replace the missing teeth. Aggressive overtreatment with implant placement in a growing child has been proven to be ill conceived in the scientific literature.[23] Preservation of existing natural elements may contribute in a positive manner to the adolescent "sense of self." The role of the prosthodontist directly participating in the activities of a multidisciplinary craniofacial team cannot be understated, and the patients will appreciate the prudent decisions for their well-being achieved by team consensus.

Acknowledgment

The prostheses fabricated for patient treatment were expertly completed by Leonard Marotta, MDT, CDT, PhD.

References

1. Shaikh R, Shusterman S: Delayed dental maturation in cleidocranial dysplasia. ASDC J Dent Child 1998;65:325-329
2. Chitayat D, Hodgkinson KA, Azouz EM: Intrafamilial variability in cleidocranial dysplasia: a three generation family. Am J Med Genet 1992;42:298-303
3. Mehta L, Verma IC: Cleidocranial dysplasia: underdiagnosed and misdiagnosed? Indian J Pediatr 1992;59:633-636
4. Siggers DC: Cleidocranial dysostosis. Dev Med Child Neurol 1975;17:522-524
5. Koch PE, Hammer WB: Cleidocranial dysostosis: review of the literature and report of case. J Oral Surg 1978;36:39-42
6. Mundlos S, Otto F, Mundlos C, et al: Mutations involving the transcription factor CBFA1 cause cleidocranial dysplasia. Cell 1997;89:773-779
7. Mundlos S: Cleidocranial dysplasia: clinical and molecular genetics. J Med Genet 1999;36:177-182
8. Cooper SC, Flaitz CM, Johnston DA, et al: A natural history of cleidocranial dysplasia. Am J Med Genet 2001;104:1-6
9. Becker A, Lustmann J, Shteyer A: Cleidocranial dysplasia: part 1–general principles of the orthodontic and surgical treatment modality. Am J Orthod Dentofac Orthop 1997;111:28-33
10. Becker A, Shteyer A, Bimstein E, et al: Cleidocranial dysplasia: part 2—treatment protocol for the orthodontic and surgical modality. Am J Orthod Dentofac Orthop 1997;111: 173-183
11. Smylski PT, Woodside DG, Harnett BE: Surgical and orthodontic treatment of cleidocranial dysostosis. Int J Oral Surgery 1974;3:380-385
12. Hall RK, Hyland AL: Combined surgical and orthodontic management of the oral abnormalities in children with cleidocranial dysplasia. Int J Oral Surg 1978;7: 267-273
13. Richardson A, Swinson T: Combined orthodontic and surgical approach to cleidocranial dysostosis. [Abstract] Trans Eur Orthod Soc 1987;63:23
14. Behlfelt K: Cleido-cranial dysplasia: diagnosis and treatment concept. [Abstract] Trans Eur Orthod Soc 1987;63:25
15. Petropoulos VC, Balshi TJ, Balshi SF, et al: Treatment of a patient with cleidocranial dysplasia using osseointegrated implants: a patient report. Int J Oral Maxillofac Implants 2004;19:282-287
16. Becker A, Shteyer A: A surgical and orthodontic approach to the dentition in cleidocranial dysostosis. [Abstract] Trans Eur Orthod Soc 1987;63:121
17. Sarwer DB, Bartlett SP, Whitaker LA, et al: Adult psychological functioning of individuals born with craniofacial anomalies. Plast Reconstr Surg 1999;103:412-418
18. Dahlquist A, Gebauer U, Ingervall B: The effect of a transpalatal arch for the correction of first molar rotation. Eur J Orthod 1996;18:257-267
19. Wise JB, Magness WB, Powers JM: Maxillary molar vertical control with the use of transpalatal arches. Am J Orthod Dentofac Orthop 1994;106:403-408
20. Riley MA, Walmsley AD, Harris IR: Magnets in prosthetic dentistry. J Prosthet Dent 2001;86:137-142
21. Kelly E, Nakamoto RY: Cleidocranial dysostosis—a prosthodontic problem. J Prosthet Dent 1974;31:518-526
22. Lombardas P, Toothaker RW: Bone grafting and osseointegrated implants in the treatment of cleidocranial dysplasia. Compend Contin Educ Dent 1997;18:509-512
23. Cronin RJ Jr., Oesterle LJ: Implant use in growing patients. treatment planning concerns. Dent Clin North Am 1998;42: 1-34