Treatment of a Patient with Cleidocranial Dysplasia Using Osseointegrated Implants: A Patient Report

Vicki C. Petropoulos, DMD, MS[1]/Thomas J. Balshi, DDS[2]/
Stephen F. Balshi, BBE[3]/Glenn J. Wolfinger, DMD[4]

This patient report describes the treatment of a 42-year-old woman with cleidocranial dysplasia. Endosseous implants were used to restore the mandibular and maxillary arches with fixed prostheses. Six implants were placed in the mandible and immediately loaded with an acrylic resin fixed prosthesis. In the maxillary arch, 10 implants were submerged for 4 months prior to functional loading. A transitional denture was relined and placed in the maxilla 10 days after implant placement. Three months later, a definitive mandibular prosthesis was fabricated. The definitive maxillary restoration was delivered 6 months after surgery. The most recent follow-up, 6 months after delivery, confirmed a satisfactory treatment result to date. INT J ORAL MAXILLOFAC IMPLANTS 2004;19:282–287

Key words: *cleidocranial dysplasia, dental prostheses, dental implants, edentulism, immediate loading, osseointegration*

Cleidocranial dysplasia is a rare inherited skeletal dysplasia. It was first described in 1897 by Marie and Sainton,[1] who termed the condition *cleidocranial dysostosis*. It has since been known as *cleidocranial dysplasia* in recognition of its underlying pathology as a generalized skeletal dysplastic condition.[2] The pattern of inheritance is usually autosomal dominant, although it has been suggested that between 20% and 40% of cases represent new mutations.[3] Best known for its dental and clavicular abnormalities, cleidocranial dysplasia is a bone disorder caused by a defect in the *CBFA1* gene of chromosome 6p21. This gene, when expressed under normal conditions, guides osteoblastic differentiation and appropriate bone formation.

Patients with cleidocranial dysplasia tend to be of short stature and have proportionally large heads with pronounced frontal and parietal bossing. They frequently have ocular hypertelorism, a broadly based nose, and a depressed nasal bridge (Figs 1a and 1b). Most dramatically, unerupted permanent teeth and supernumerary teeth are sometimes found (Fig 1c).

Treatment of the dental problems associated with cleidocranial dysplasia may be difficult.[4] Therapeutic options include extraction of all teeth followed by the fabrication of dentures or a crown sleeve coping overdenture,[5] autotransplantation[6] of selected impacted teeth followed by prosthetic restoration, or removal of primary and supernumerary teeth followed by exposure of permanent teeth that are subsequently extruded orthodontically. The use of implants in a patient with cleidocranial dysplasia to support a removable overdenture has been documented.[7]

[1]Consultant Prosthodontist, VA Medical Center, Philadelphia, Pennsylvania; Assistant Professor, Department of Restorative Dentistry, University of Pennsylvania School of Dental Medicine, Philadelphia, Pennsylvania.
[2]Prosthodontics Intermedica, Institute for Facial Esthetics, Fort Washington, Pennsylvania.
[3]Director of Biomedical Engineering, Prosthodontics Intermedica, Institute for Facial Esthetics, Fort Washington, Pennsylvania; Postgraduate Student, Drexel University, School of Biomedical Engineering and Science, Philadelphia, Pennsylvania.
[4]Prosthodontics Intermedica, Institute for Facial Esthetics, Fort Washington, Pennsylvania; Staff Prosthodontist, VA Medical Center, Philadelphia, Pennsylvania; Lecturer, Department of Prosthodontics, Columbia University School of Dental and Oral Surgery, New York, New York.

Correspondence to: Dr Vicki C. Petropoulos, 4001 Spruce Street, University of Pennsylvania School of Dental Medicine, Department of Restorative Dentistry, Philadelphia, PA 19104-6003. Fax: +215 573 9606. E-mail: vpetropoulos@aol.com.

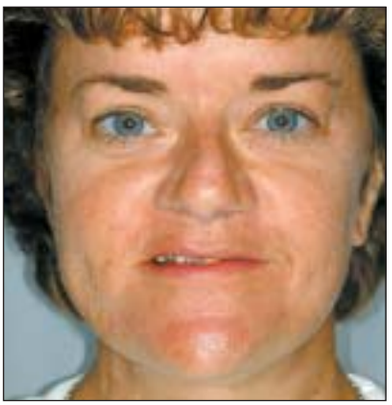


Fig 1a Preoperative full face showing. Note the broad base of the nose and the depressed nasal bridge.

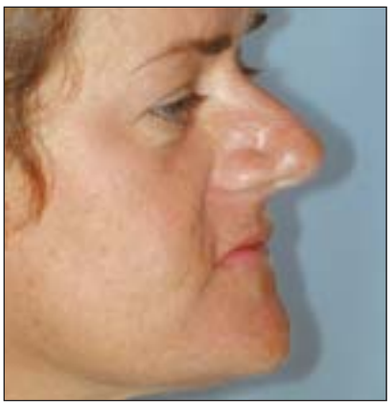


Fig 1b Preoperative profile view of patient.

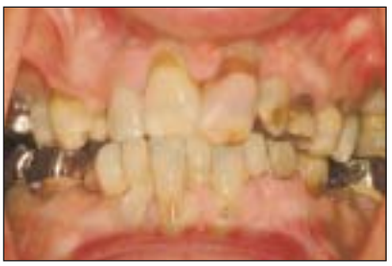
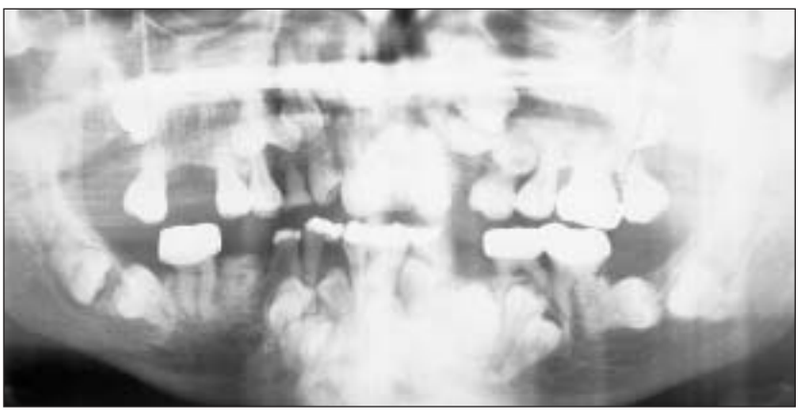


Fig 1c Preoperative intraoral photograph of maxillary and mandibular teeth in occlusion.

Fig 2 Panoramic radiograph showing numerous supernumerary and primary teeth.



However, there is a paucity of documented cases using implants to support an implant-supported fixed prosthesis with this population. Likewise, immediate loading and function have not been studied with this population. Although cleidocranial dysplasia is a bone disorder caused by a defect in the gene that guides osteoblastic differentiation and bone formation, using implants in such a case seems logical since there have been documented cases of bone formation around orthodontically erupted teeth in patients with cleidocranial dysplasia.[8]

CASE REPORT

Patient History

The patient was a 42-year-old woman born with cleidocranial dysplasia. She was missing a piece of her clavicle and had the facial anomalies common among those with this condition (Figs 1a and 1b). The patient, who was in good general health, with no known allergies or sensitivities to medications, presented for treatment related to reconstruction of her dentition. As a child, she had undergone several unsuccessful surgeries to expose unerupted teeth. Her chief complaints were, "I have ugly teeth," "I am unable to chew properly," and "I want to hide." Throughout her life, she had been self-conscious about the appearance of her mouth (Fig 1c) and was not comfortable talking or eating with people.

Clinical Evaluation and Diagnosis

At the initial visit, the patient presented with the following teeth in her maxilla: 2(17), 3(16), 4(15), 8(11), 9(21), 10(22), 14(26), and 15(27). She had the following teeth in her mandible: 19(36), 20(35), 21(34), 22(33), 24(31), 25(41), 26(42), and 29(45). Comprehensive clinical and radiographic examinations were performed. Lateral, cephalometric, and panoramic radiographs (Fig 2) revealed that in addition to the aforementioned teeth, the patient had 29 supernumerary teeth (14 maxillary, 15 mandibular; Table 1) and 4 primary teeth (1 maxillary, 3 mandibular). Diagnostic casts were articulated at an improved occlusal vertical dimension, permitting laboratory technicians to fabricate provisional dentures.

Table 1 Location of Supernumerary Teeth

Region	No. of teeth
Maxilla	
Right	
Third molar	2
Second molar	1
Second premolar	1
First premolar	1
Canine	2
Lateral incisor	1
Left	
Canine	2
First premolar	2
Second premolar	1
Third premolar	1
Mandible	
Left	
Third molar	1
Second molar	1
Second premolar	1
First premolar	1
Canine	1
Lateral incisor	1
Central incisor	1
Right	
Central incisor	1
Lateral incisor	1
Canine	1
First premolar	1
Second premolar	1
First molar	1
Second molar	1
Third molar	1

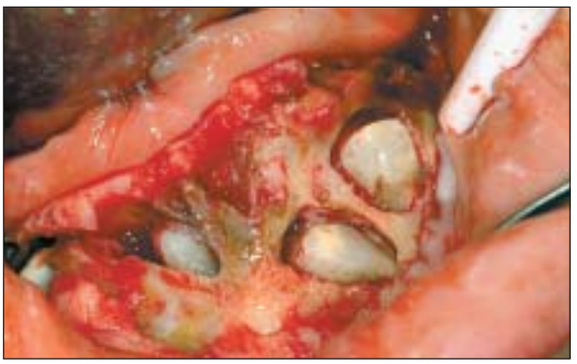


Fig 3a Encapsulated maxillary supernumerary teeth.

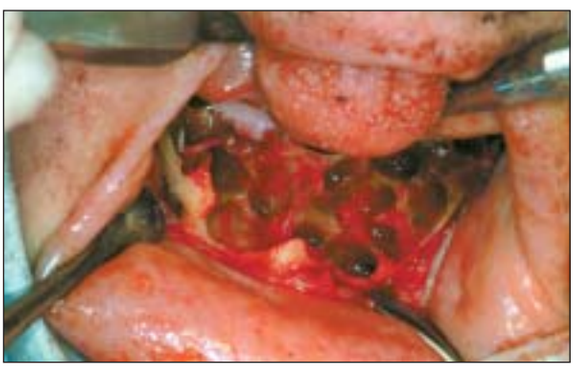


Fig 3b Large voids in the alveolar bone following extractions.

Tooth Extraction

General anesthesia, propofol (Diprivan; AstraZeneca, Wilmington, DE) as an induction agent followed by isofluorane (Florane; Abbott Laboratories, Dallas, TX) for maintenance of anesthesia were administered by a board-certified anesthesiologist using nasal intubation. This was followed by local anesthesia, 18 mL of 0.5% Marcaine (Abbott Laboratories) with 1:200,000 epinephrine. All the patient's teeth were extracted. Curettes were used to remove the soft tissue encapsulation from the deeply impacted supernumerary teeth (Fig 3a). Following the extractions, alveoloplasty was used to harvest bone that was then regrafted into the osseous defects (Fig 3b). The grafted bone was mixed with a tetracycline solution (Ivax Pharmaceuticals, Miami, FL) and loosely packed. Primary closure of the flaps created a biologic seal immediately prior to the relining of the provisional removable prostheses. The patient returned for suture removal and monthly relining of the provisional prostheses using a temporary denture retaining material (Visco-Gel; Dentsply, York, PA).

Surgical Placement of Implants in the Mandible

Three months after the extractions, the patient presented for the placement of dental implants. General anesthesia was administered by a board-certified anesthesiologist using nasal intubation, followed by local anesthesia, using the same drugs used for tooth extraction. In the mandibular arch, a crestal incision with dissection and flap elevation was made bilaterally from second molar to second molar. Six 3.75 × 13-mm implants (Brånemark TiUnite Mk III; Nobel Biocare, Göteborg, Sweden) were placed in the left first premolar, left canine, left central incisor, right central incisor, right lateral incisor, and right canine regions. All mandibular implants were immediately loaded with abutments and an acrylic resin fixed prosthesis (Figs 4a and 4b).[9]

Surgical Placement of Implants in the Maxilla

Immediately following placement of the mandibular implants, 10 implants were placed in the maxilla following the 2-stage Brånemark protocol.[10] Five 4 × 15-mm implants were placed in the right third

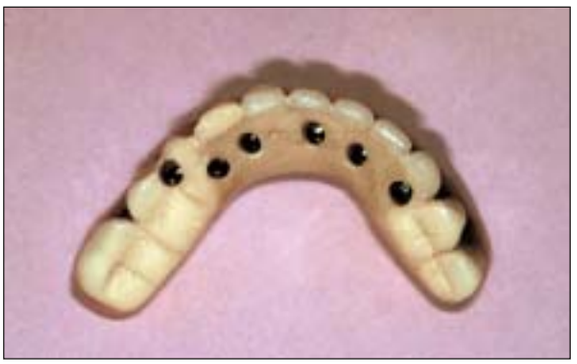


Fig 4a Occlusal view of the mandibular temporary prosthesis.

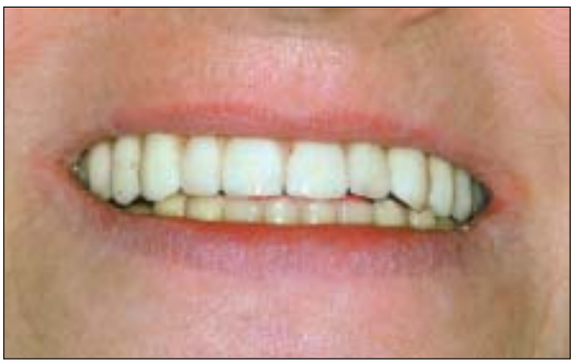


Fig 4b Temporary mandibular prosthesis in place with transitional complete maxillary denture.

molar, right first molar, right central incisor, left second molar, and left third molar regions, and five 4 × 13-mm implants were placed in the right canine, right lateral incisor, left central incisor, left canine, and left first premolar regions (Brånemark TiUnite Mk IV; Nobel Biocare). Primary closure was established using Vicryl sutures (Ethicon, Somerville, NJ). The maxillary provisional denture was relined and seated 10 days after the implant surgery.

Postsurgical Care

Following the surgery, the patient was provided with postsurgical instructions, cold therapy, standard medications (anti-inflammatory pain medication, steroids to control swelling, antibiotics, and chlorhexidine rinse), and dietary restrictions, which included a strictly soft diet for 8 weeks.

Definitive Prosthesis for the Mandible

Three months after surgical and restorative procedures for the mandibular arch, the patient presented for fabrication of the definitive prosthesis for the mandibular arch. An interocclusal registration was made using vinyl polysiloxane bite registration material (Regisil; Dentsply). The final impression was made using the existing mandibular fixed prosthesis as an impression vehicle. Heavy body vinyl polysiloxane impression material (Reprosil; Dentsply) was inserted beneath the prosthesis using a syringe, and a pickup impression was made. A master cast was created by placing abutment analogs of the modified impression copings within the fixed prosthesis. In the authors' experience, use of the immediately loaded fixed prosthesis as an impression vehicle creates an exceptionally accurate master cast. The maxillary denture was duplicated using alginate impression material. The interocclusal registration and the provisional restorations were used to articulate the maxillary edentulous cast against the mandibular master cast. The laboratory then began fabrication of the definitive metal-reinforced mandibular prosthesis.

The patient participated in try-ins for functional and esthetic assessment as well as verification of the recorded vertical dimension of occlusion. The definitive prosthesis was then delivered.

Fabrication of a Temporary Prosthesis for the Maxilla

Six months after surgical placement, the maxillary implants were uncovered. All the implants were osseointegrated except for the one in the right third molar region, which was encapsulated in fibrous tissue. This implant was removed. The abutments were connected, and a panoramic radiograph was taken to verify the fit. An acrylic resin temporary prosthesis was fabricated.

Definitive Prosthesis for the Maxilla

The final impression for the definitive prosthesis was made using fast-setting plaster (Kerr, Romulus, MI) to block out the undercut areas. The maxillomandibular relationship was recorded using Regisil at the existing vertical dimension of occlusion.

The gold-cast framework was tried-in 2 weeks after the final impressions. A panoramic radiograph was taken to verify the fit. It was necessary to section the casting. A pickup impression was made using Reprosil impression material along with another interocclusal record. The impression was poured in the laboratory and articulated.

The definitive prosthesis was fabricated of porcelain fused to gold. A panoramic radiograph was taken to verify the fit. The occlusion was adjusted so that all the contacts were even. The access holes were sealed using cotton and elastic light-curing material (Fermit; Ivoclar Vivadent, Schaan Lichtenstein). The patient was extremely pleased with the results (Figs 5a to 5c).

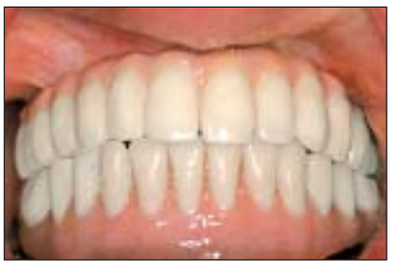


Fig 5a *(Left)* Maxillary porcelain-fused-to-gold restoration opposing mandibular gold bar denture teeth.

Fig 5b *(Right)* Postoperative esthetics.

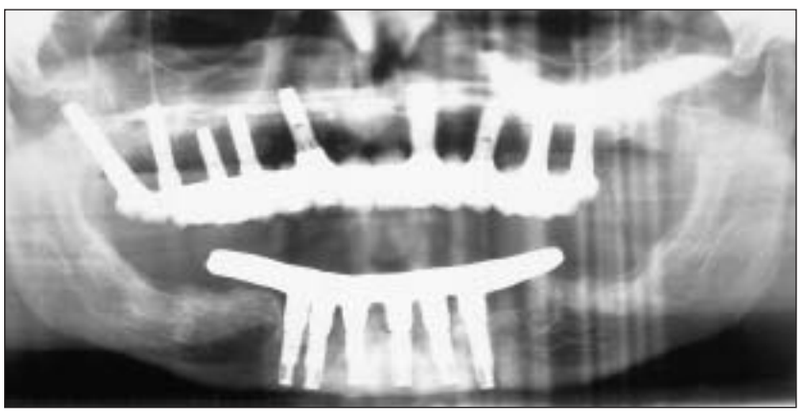


Fig 5c A postoperative panoramic radiograph.

DISCUSSION

This article describes the use of endosseous implants in treating a 42-year-old woman with cleidocranial dysplasia. The described protocol offers an effective treatment option for patients with cleidocranial dysplasia and eliminates the long-standing struggle with ill-fitting, uncomfortable, or unsightly removable prostheses. It relies heavily on coordinated surgical and restorative treatment provided by an experienced prosthodontic team. The entire reconstruction took 11 months from the time the patient first presented at the Prosthodontics Intermedica Dental Implant Center. The benefits of providing therapy in a timely fashion cannot be overstated.

Despite a lack of evidence-based data to support the potential for ossoeointegration around titanium implants in a patient with cleidocranial dysplasia, there was evidence that bone remodeling and osseointegration occurred with this patient despite the fact that this genetic defect affects osteoblastic activity. Therefore, it may be concluded based on this patient report that osseointegration had effectively stabilized the implants. For a more definitive understanding of the specific biologic and biochemical mechanisms involved in cleidocranial dysplasia, long-term studies are needed. Although the favorable outcome with this individual patient demonstrates the potential for successful management of similar congenital anomalies, additional clinical research is necessary for universal application.

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REFERENCES

1. Marie P, Sainton P. Observation d'hydrocephailie hereditaire (pere et fils) par vice de development du crane et du cerveux. Bull Soc Med Hop Paris 1897;14:706–712.
2. Butterworth C. Cleidocranial dysplasia: Modern concepts of treatment and a report of an orthodontic resistant case requiring a restorative solution. Dent Update 1999;12:458–463.
3. Fitchet SM. Cleidocranial dysostosis: Hereditary and familial. J Bone Joint Surg 1929;11:833–866.
4. Shafer WG, Hine MK, Levy BM. Diseases of the bone and joints. In: Shafer WG, Hine MK, Levy BM (eds). A Textbook of Oral Pathology. Philadelphia: Saunders, 1974:622–664.
5. Weintraub GS, Yalisove IL. Prosthodontic therapy for cleidocranial dysostosis: Report of a case. J Am Dent Assoc 1978;96:301–305.
6. Jensen BL, Kreiborg S. Dental treatment strategies in cleidocranial dysplasia. Br Dent J 1992;172:243–247.
7. Lombardas P, Toothaker RW. Bone grafting and osseointegrated implants in the treatment of cleidocranial dysplasia. Compend Contin Educ Dent 1997;18:509–514.
8. 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.
9. Balshi TJ. The Biotes conversion prosthesis: A provisional fixed prosthesis supported by osseointegrated titanium implants for restoration of the edentulous jaw. Quintessence Int 1985;16:667–677.
10. Adell R, Lekholm U, Brånemark P-I. Surgical procedures. In: Brånemark P-I, Zarb GA, Albrektsson T (eds). Tissue-Integrated Prostheses: Osseointegration in Clinical Dentistry. Chicago: Quintessence, 1985:199–220.