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Platform switching in the treatment of Cleidocranial Dysplasia: a case report

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ABSTRACT

Background Cleidocranial dysplasia is a very rare occurrence, its incidence being 1: 1,000,000.
Case report This report describes the treatment of a 31-year-old woman with cleidocranial dysplasia treated with expanded platform implants. All mandibular and maxillary teeth were extracted and porcine collagenized bone was used to cover the bone defects in both arches. Six months later, four expanded platform implants were used to restore the mandibular arch, but one of them failed before the prosthesis was placed. In the maxillary arch a complete denture was relined and placed in the maxilla. The definitive mandibular restoration was delivered 3 months after surgery.
Conclusion Since early diagnosis of cleidocranial dysplasia is essential for choosing the appropriate treatment approach, clinicians should be aware of its characteristic features.

KEYWORDS Cleidocranial dysplasia; Dental implants; Dental prosthesis; Edentulism.

INTRODUCTION

Cleidocranial dysplasia (CCD) is a rare autosomal condition affecting bones, generally the calvarian but also the clavicular bone, that undergo intramembranous ossification.
CCD was first described by Pierre Marie and Paul Sainton in 1897 (1), since then, over 1000 cases have been documented in the medical literature, which termed the condition cleidocranial disostosis (2). It has since been known as CCD 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). The disorder is caused by mutation in the CBFA1 gene, on the short arm of chromosome 6p21 (3). The prevalence of cleidocranial dysostosis is estimated one per million, without sex or ethnic group predilection (4).
Patients with CCD tend to be of short stature and have proportionally large heads with pronounced frontal and parietal bossing of the skull. They frequently have ocular hypertelorism, a broadly based nose, and a depressed nasal bridge. The ability to approximate the shoulders anteriorly is related to clavicular hypoplasia and is the classic diagnostic sign of the disorder (5). Moreover, unerupted permanent teeth and supernumerary teeth are sometimes found. Underdevelopment of the maxilla and relative mandibular prognathism are common. Prolonged exfoliation of the primary dentition, unerupted supernumerary teeth, and the irregularly and partially erupted secondary dentition result in occlusal anomalies. The presence of the second permanent molars together with the primary dentition and wide spacing in the lower incisor area are typical dental signs (5).
The dental abnormalities associated with it present a remarkable challenge in 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 several surgeries might be required. There are many difficulties in the early diagnosis of CCD because most of the craniofacial abnormalities become obvious only during adolescence (5).
Treatment options for the management of impacted teeth are separated into four categories: observation, intervention, relocation and extraction (6). In regions where no supernumerary teeth are formed, eruption may

also be improved by removal of the primary teeth and surgical exposure of the underlying permanent teeth. Conventional orthodontic treatment and eventually autotransplantation of teeth may still be necessary in the future, but it can be anticipated that the new strategy, with much earlier intervention, can reduce the extent of surgical and orthodontic interventions, which have previously been of extremely long duration, uncomfortable to patients and often of limited success (7). Therapeutic options include extraction of all teeth followed by the fabrication of dentures or a crown sleeve coping overdenture (8), autotransplantation (7) 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 CCD to support a removable overdenture has been documented (9). However, there is a paucity of documented cases using implants to support a fixed prosthesis with this population. Likewise, immediate loading and function have not been studied in these subjects. Although CCD is a bone disorder caused by a defect in the gene that guides osteoblastic differentiation and bone formation, the use of implants in such cases seems logical, since there have been documented cases of bone formation around orthodontically erupted teeth in patients with CCD (10).

The aim of this article is to report a rare case of a woman with CCD who was restored in the mandibular and maxillary arches with endosseous implants supporting fixed prostheses.

CASE REPORT

Patient history

A 31-year-old woman with a history of CCD originally presented to the General and Implant Dentistry Department, Faculty of Medicine and Dentistry, University of Murcia (Spain) with the chief complaint of an ill-fitting mandibular and maxillary partial denture. She was missing both clavicles and had the facial anomalies typical of this condition. The patient was in good general health, with no known allergies or sensitivities to medications. Throughout her life, she had been self-conscious about the appearance of her mouth and could not comfortably eat or talk with peop1e.

Clinical evaluation and diagnosis

Examination of the oral cavity revealed multiple over-retained permanent teeth and one supernumerary tooth, particularly in the anterior maxilla and mandible on the right and left side (Fig. 1).

On evaluating the panoramic radiograph, the classical signs of CCD were immediately recognized (Fig. 2). The patient had 42 teeth in her jaws. Some of the teeth were erupted but most of them were unerupted and the supernumerary tooth mimicked a premolar in shape. Gonial angles on both sides of the mandible were missing and maxillary sinuses were underdeveloped.

At the initial visit, on the radiograph the patient presented with the following teeth in her maxilla: four primary teeth (55-53-63-65), two permanent teeth erupted (16-26) and 14 retained teeth (11-12-13-13-15-16-17-18,21-22-23-24-25-27-28). She had the following teeth in her mandib1e: five primary teeth (74-73-72-82-83), one permanent erupted tooth (35) and 15 retained teeth (38-37-36-34-33-32-31-41-42-43-44-45-46-47-48). The patient had 1 supernumerary tooth in the right side of the mandible.

Comprehensive clinical and radiographic examinations were performed. Diagnostic casts were articulated at an improved occlusal vertical dimension, permitting laboratory technicians to fabricate provisional dentures.

Teeth extractions

All the patient's teeth were extracted (Fig. 3, 4).

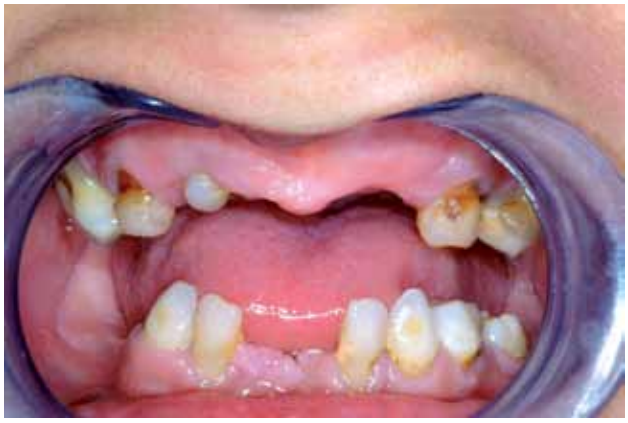

FIG. 1 Clinical aspect of the oral cavity of the patient.

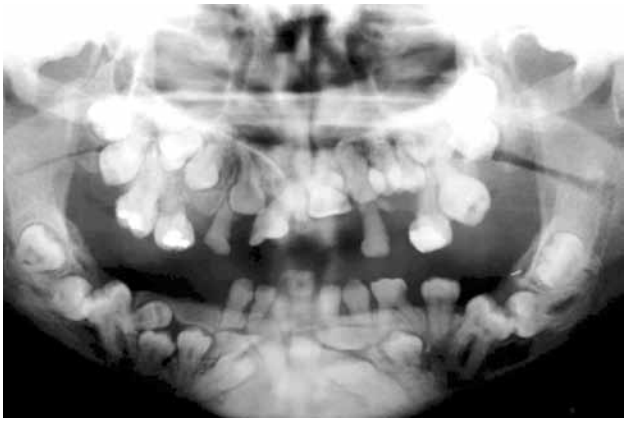

FIG. 2 Panoramic radiograph: the classical oral signs of cleidocranial dysplasia could be observed.

Following the extractions, alveoloplasty was used to harvest bone that was then regrafted mixed with collagenized porcine bone (MP3, Tecnoss dental, Pianezza, TO, Italy) into the osseous defects and covered with collagenized membrane (Fig. 5). 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 fifteen days after the surgery and relining of the provisional prostheses using a temporary

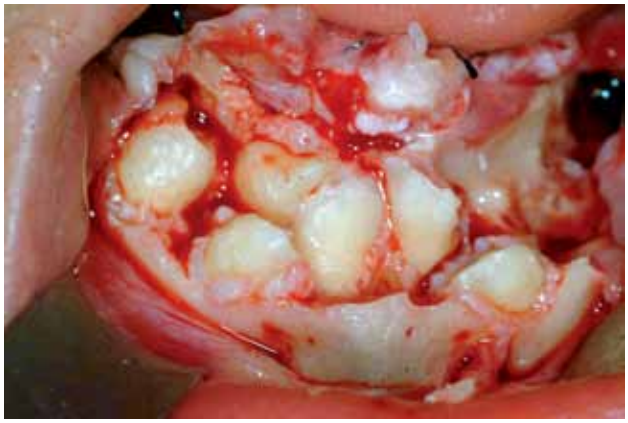

FIG. 3 Mandibular arch before teeth extractions.

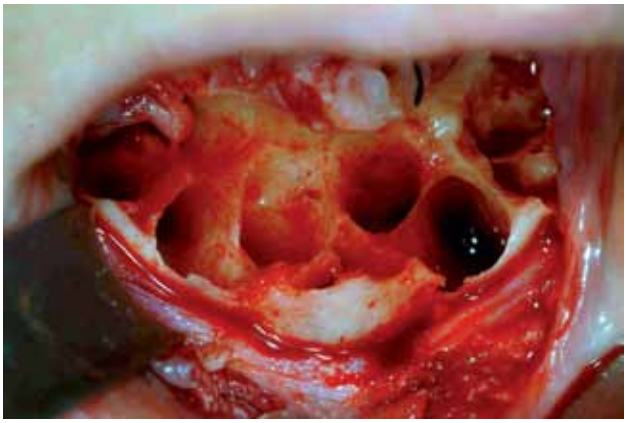

FIG. 4 Mandibular aspects following teeth extractions.

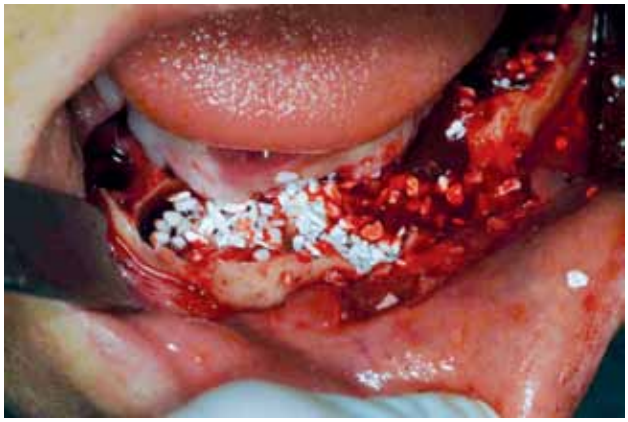

FIG. 5 Grafting of autologous bone mixed with collagenized porcine bone into the bone defects.

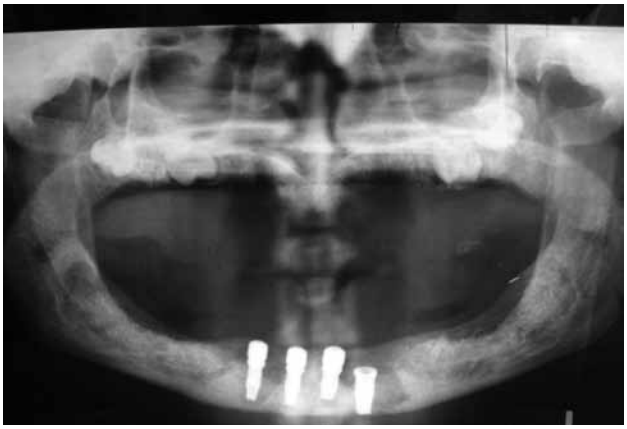

FIG. 6 Panoramic radiograph following mandibular implants placement.

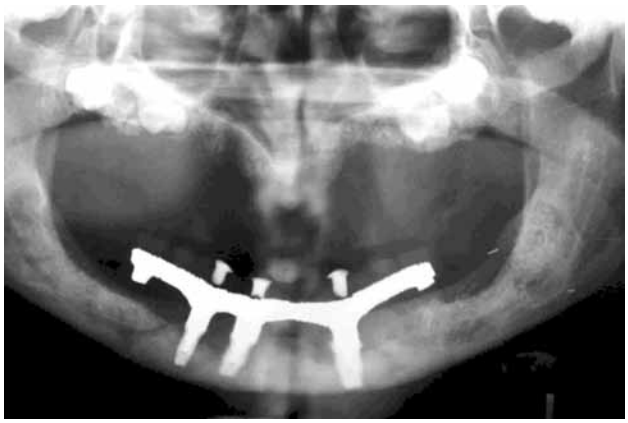

FIG. 7 Radiographic aspect of the metal-reinforced mandibular prosthesis.

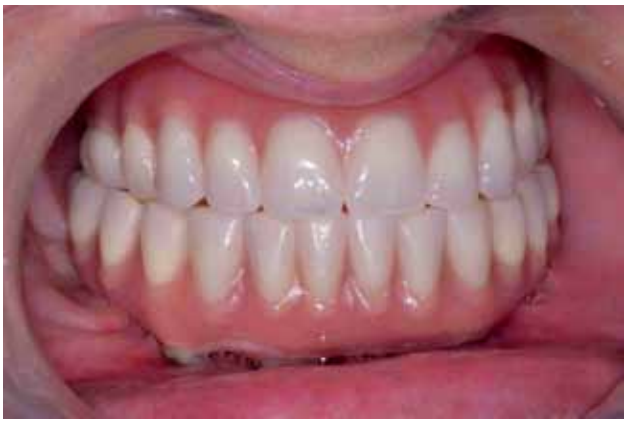

FIG. 8 Clinical view of the delivered prosthesis.

denture retaining material (Ufi Gel SC, VOCO, GMBH, Cuxhaven, Germany).

Surgical placement of implants in the mandible

Six months after the extractions, the patient presented for the placement of dental implants in the mandible. Local anesthesia was administered and a mandibular arch crestal incision was made bilaterally from second molar to second molar. A biopsy was taken from the grafted area to be analyzed. Three 4/5/4-mm implants (Biomet 3i, Palm Beach Gardens, FL, USA) and one 5/6/5 mm implant were placed in the front area between both mental nerves (Fig. 6). Following the surgery, the patient was provided with postsurgical instructions, namely cold therapy, standard medications (anti-inflammatory pain medication, steroids to control swelling, antibiotics, and chlorhexidine rinse), and diet restrictions, which included a strictly soft diet for 8 weeks.

Definitive prosthesis for the mandible

Three months after surgery in the mandibular arch, the final impression was made using heavy body/light body polyether impression material (Impregum Penta, 3M, St.Paul, MN, USA). A master cast was created by placing abutment analogs of the modified impression copings within the fixed prosthesis. The maxillary denture was made using alginate impression material. The interocclusal registration and the provisional restorations were used to articulate the maxillary conventional denture against the mandibular master cast. The laboratory then began fabrication of the definitive metal-reinforced mandibular prosthesis (Fig. 7). The definitive prosthesis was then delivered after 15 days (Fig. 8).

DISCUSSION AND CONCLUSION

In the literature cases reporting the use of expanded platform implants in patients affected by CCD are rare. The described protocol offers an effective treatment option for patients with CCD and eliminates the long-standing struggle with ill-fitting, uncomfortable, or unsightly removable prostheses. The entire reconstruction took 9 months from the time the patient first presented at the General and Implant Dentistry Department, Faculty of Medicine and Dentistry, University of Murcia (Spain).

The radiographic evaluation of patients is the most important and reliable means to confirm the diagnosis, since radiological findings of CCD are pathognomonic, i.e. broad sutures, large fontanels persisting into adulthood, numerous wormian bones and unerupted teeth (1, 3). Despite a lack of evidence-based data to support the potential for ossoeointegration around titanium implants in a patient with CCD, there was evidence that bone remodeling and osseointegration occurred in this patient despite the fact that this genetic defect affects osteoblastic activity (9).

For a more definitive understanding of the specific biologic and biochemical mechanisms involved in CCD, long-term studies are needed. Although the favorable outcome with this individual patient demonstrates the potentially successful management of similar congenital anomalies, additional clinical research is necessary for universal application. Therefore, based on this patient report, it may be concluded that osseointegration had effectively stabilized the implants.

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