Objective. The aim of this study was to describe the rehabilitation of patients diagnosed with recessive dystrophic epidermolysis bullosa (RDEB), with edentulous maxillas and/or mandibles, by using fixed full-arch short-expand prostheses supported by 4 anterior implants.

Study design. A retrospective study was carried out to study edentulous patients with RDEB rehabilitated with fixed full-arch prostheses supported by 4 anterior implants.

Results. In total, 32 anterior implants were placed and used to support 8 full-arch fixed prostheses; 20 implants were placed in the maxilla by using osteotomes and 12 in the mandible by using conventional drilling. Implant success rate was 100% after an average follow-up of 22.9 (range 12-48) months after prosthetic loading. Patient satisfaction with the implant therapy was very high (mean 9.0) for all the factors assessed.

Conclusions. Fixed full-arch short-expand prostheses supported by 4 anterior implants can be successfully used to rehabilitate patients with recessive dystrophic epidermolysis bullosa, considerably improving these patients’ quality of life. (Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2011;112:e4-e10)

Epidermolysis bullosa (EB) is a group of disorders characterized by the mechanical fragility of the skin which recurrently develops blisters and vesicles. Three major presentations (simplex, junctional, and dystrophic) and 25 subtypes of epidermolysis bullosa exist, the recessive dystrophic form being the presentation in which the oral mucosa is more severely affected.

Systemic features of EB include blisters all over the body, especially in areas of friction, such as hands, feet, elbows, and knees, which break leaving painful ulcerations that often heal with soft tissue contraction (Fig. 1, A). In the most severe forms, junctional and dystrophic EB, tissue contraction frequently causes digit syndactyly, which leads to stump formation, and stenosis of the upper third of the esophagus, which originates dysphagia. Oral features include repeated occurrence of blisters, erosions, and scars, which lead to limited mouth opening, ankyloglossia, and elimination of buccal sulci; alveolar bone resorption and atrophy of the maxillaries; and increased risk of oral carcinoma. Routine dental care or even normal tooth brushing might cause bullae on the oral mucosa.

Oral rehabilitation in these patients with implant-supported fixed prostheses has proved to be a treatment with high success rates that provides significant improvements to their quality of life. However, implant treatment is hindered by the soft-tissue fragility, the limited oral opening, and the severe maxillary atrophy. Extreme care during the surgery allows overcoming soft-tissue fragility; the use of short-expand full-arch prostheses supported by 4 anterior implants may allow overcoming the limited oral opening and bone atrophy, thereby providing edentulous patients with recessive dystrophic epidermolysis bullosa (RDEB) with a fixed prosthetic solution. Residual bone between the mental foramina (in the mandible) and in the canine buttress or toward the palatal (in the maxilla) allows the placement of dental implants. Treatment of atrophic maxillaries by using fixed prostheses over 4 implants, frequently tilted to take advantage of residual bone, has been shown to be successful in healthy patients. Regarding patients with EB, a few cases of rehabilitations with overdentures and fixed full arch supported by 4 implants have been reported (Table I).

The aim of the present clinical report was to describe the rehabilitation of patients diagnosed with RDEB, with edentulous maxillas and/or mandibles, by using fixed full arch short-expand prostheses supported by 4 anterior implants.
MATERIALS AND METHODS

Fifteen patients with RDEB presenting for dental treatment in a university dental school were treated with dental implants between January 2005 and December 2010; a retrospective study was carried out to study edentulous patients with RDEB rehabilitated with fixed full-arch prostheses supported by 4 anterior implants. The inclusion criteria were: 1) a diagnosis of RDEB; 2) completely edentulous maxilla and/or mandible, or partially edentulous with indication of extraction of remaining teeth; 3) acceptance of implant treatment; 4) rehabilitation with full-arch fixed prosthesis supported by 4 anterior implant; and 5) a minimum follow-up of 1 year after implant loading. Patients with incomplete protocols were excluded. Of the 15 patients, 6 met the requirements to be included in this study. Three patients were not completely edentulous and were rehabilitated with partial fixed prostheses, 4 patients received <4 implants and were rehabilitated with removable prostheses, 1 patient received 5 implants, and 1 patient had been followed for <12 months.

Microstomia was severe in all cases, according to the classification of Naylors et al.\textsuperscript{15} (Fig. 1, B), and in all cases severe ankyloglossia (i.e., adherence of the

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Fig. 1. (Continued).
Fig. 1. (Continued).
tongue to the floor of the mouth) and severe obliteration of the oral vestibule were also found. All patients included in the study exhibited marked intraoral involvement, with devastating alterations in the soft and hard tissues, and all had antecedents of bleeding blisters, rampant dental caries, and loss of dentition (Fig. 1, C and D). Panoramic radiographs and computerized tomograms of all patients were obtained and studied (Fig. 1, E-G).

All surgeries were carried out under local anesthesia (4% articaine and adrenaline 1:100,000; Ultracain; Aventis Pharma, Bad Soden, Germany). The anesthetic solution was injected deeply into the tissues and at a sufficiently slow rate to prevent tissue distortion. The patients’ lips were lubricated with petroleum jelly to avoid tissue friction or irritation of the mucosa and bulla formation. In all cases, conscious sedation was performed with 1% propofol solution (Diprivan; AstraZeneca Pharma, Madrid, Spain) administered by an anesthesiologist.

In the maxilla, implant sites were created by using osteotomes, because conventional drilling procedure would have destroyed the entire residual alveolar process, thus complicating primary stability of the implants. In contrast, in the mandible drills were used. The minimum necessary irrigation with sterile saline solution was used to minimize the use of aspiration, which can traumatize soft tissues. Whenever necessary, aspiration was done in contact with bone, not with soft tissues. Defcon Avantblast TSA surface (Impladent; Sentmenat, Barcelona, Spain) and ITI SLA surface (Straumann Institute, Waldenburg, Switzerland) implants were used. Peri-implant defects were regenerated with particulated autologous bone graft collected from the surgical drills during implant bed preparation, or with tricalcium betaphosphate Kera-Os (Keramat, Coruña, Spain) in cases in which autogenous bone was not available because implant sites had been created by using osteotomes. Collagen resorbable membranes (Lyostypt; B. Braun, Aesculap, Tuttlingen, Germany) were used to protect the particulate bone grafts (Fig. 1, H-V).

Panoramic radiographs were obtained immediately after surgery (digital orthopantomograph Op 100; Instrumentarium Imaging, Tuusula, Finland). Oral antibiotics (Clamoxyl; Glaxo-Smith-Kline; 500 mg every 8 hours for 7 days) and nonsteroidal antiinflammatory medication (ibuprofen; Bexistar; Laboratorio Bacino; 600 mg every 8 hours for 3 days) were prescribed. Sutures were removed 7 days after the surgery. The implants were allowed to integrate for 3 months in the mandible and 6 months in the maxilla before prosthetic loading. Patients were then rehabilitated with short-expand fixed full-arch prostheses (Fig. 1, W-Z).

Patients were clinically monitored 1 and 3 months after the surgery, and every 6 months thereafter; control panoramic radiographs were obtained every 12 months. The definition of implant success was based on Albrektsson et al.’s clinical and radiologic criteria: 1) absence of clinically detectable implant mobility; 2) absence of exudates, persistent inflammation, patient discomfort, or bleeding; 3) absence of periapical radiolucencies; and 4) absence of progressive bone loss >0.2 mm after the first year of implant placement. Peri-implant radiolucencies and bone loss had to be studied in panoramic radiographs, because no intraoral radiographs could be taken owing to the extreme fragility of the oral mucosa.

Six months after prosthetic restoration, patients were asked to complete a questionnaire measuring their satisfaction and the psychologic impact of their oral health status. Comfort and retention, function, esthetics and appearance, taste, speech, and self-esteem were assessed. The questionnaire was carefully explained to the patients, and any doubts were resolved before asking them to place a mark on a visual analog scale (VAS) corresponding to their level or satisfaction or discontent with each factor. The VAS was a horizontal beam 10 cm in length, with the left end representing 0% (the negative limit) and the right end representing 100% (the positive limit).

This research was exempt from approval by the local Institutional Review Board. Each of the patients gave

<table>
<thead>
<tr>
<th>Author (year)</th>
<th>Case</th>
<th>Location</th>
<th>No. of implants placed</th>
<th>Failures</th>
<th>Follow-up (y)</th>
<th>Type of prostheses</th>
<th>% success</th>
</tr>
</thead>
<tbody>
<tr>
<td>Penarrocha (2000)</td>
<td>1</td>
<td>Maxilla</td>
<td>4</td>
<td>0</td>
<td>1-4</td>
<td>Overdentures</td>
<td>100</td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>Mandible</td>
<td>4</td>
<td>0</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lee (2007)</td>
<td>3</td>
<td>Maxilla</td>
<td>4</td>
<td>0</td>
<td>4</td>
<td>Fixed</td>
<td>100</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Mandible</td>
<td>4</td>
<td>0</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Penarrocha (2008)</td>
<td>4</td>
<td>Mandible</td>
<td>4</td>
<td>0</td>
<td>1-5</td>
<td>Fixed</td>
<td>100</td>
</tr>
<tr>
<td></td>
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<td>4</td>
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<td></td>
<td></td>
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</tr>
<tr>
<td></td>
<td>6</td>
<td>Mandible</td>
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<td>0</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Muller (2010)</td>
<td>7</td>
<td>Maxilla</td>
<td>4</td>
<td>0</td>
<td>4-5</td>
<td>Fixed</td>
<td>100</td>
</tr>
</tbody>
</table>
RESULTS
Six patients (4 women and 2 men) diagnosed with RDEB were rehabilitated with fixed full arch prostheses supported by 4 implants; 3 patients were treated of the maxilla and the mandible, 2 of only the maxilla and 1 of only the mandible. In total, 32 anterior implants (20 Defcon and 12 ITI) were placed, 20 in the maxilla by using osteotomes and 12 in the mandible by using conventional drilling. In 16 implants, particulate bone graft (autologous or tricalcium betaphosphate Kera-Os) and resorbable collagen membranes (Lyostypt) were used to cover dehiscences or fenestrations. Those implants in which bone regeneration was performed were left submerged. Intraoperative blister complications were recorded in all cases (Fig. 1, Z); however, the postoperative course was normal in all cases. Characteristics of the patient sample are detailed in Table II.

Sutures were removed 1 week after the surgery, and patients were clinically evaluated after 1 month and 3 months and then every 6-months. The mucosa remained in good condition around all of the implants, with no peri-implant bullae being observed. Radiographic follow-up was performed every 12 months and demonstrated correct bone integration of the implants, with no significant peri-implant bone loss. The implant success rate was 100% after an average follow-up of 22.9 (range 12-48) months after prosthetic loading.

Before prosthesis placement, the patients were unable to chew, and had to swallow food in puree form to avoid esophageal ulcerations. After prosthetic rehabilitation, all patients reported being able to chew and swallow a well ground food bolus. Patient satisfaction with the implant therapy was very high (mean 9.0) for all of the factors assessed (comfort and retention, function, esthetics and appearance, taste, speech, and self-esteem).

DISCUSSION
The rehabilitation with implant-supported fixed prostheses of edentulous patients with RDEB has been shown to be a successful and excellently tolerated treatment. It allows these patients to chew food more efficiently than conventional removable prostheses, thus reducing the risk of damage to the oral and esophageal mucosae and improving their nutrition.6,7 Several authors have reported dental implant treatment in patients with EB under general anesthesia, 13,14,17 whereas others prefer local anesthesia to avoid the risk of ulcerations by intubation.6,7,17,18 In the present study, all patients were collaborative adults and surgeries could be performed under conscious venous sedation and local anesthesia.

Severe bone atrophy is one of the oral characteristics of EB which limits implant treatment.4 The osteotome technique described by Summers19 allows conservation and compaction of the residual bone, thus achieving primary fixation of implants even in atrophic maxillas. In the present study, maxillary implants were placed by using osteotomes, because conventional drilling would have destroyed the residual bony process. In the harder bone of the mandible, the conventional drilling procedure was used with the minimum irrigation necessary, because aspiration can cause tissue damage and bullae formation;7 lubricating the patient’s lips and any other tissues susceptible to contact also reduces tissue damage caused by shear forces.13,20,21

In the literature reviewed,2,12-14 a total of 7 patients diagnosed with EB had been treated with either overdentures or fixed full-arch prostheses supported by 4 implants; in total, these patients received 32 implants (12 in the maxilla and 20 in the mandible), all of which survived ≥1 year after implant placement. In healthy patients, large case series have been reported of patients treated with fixed prostheses over 4 implants.8-11 Maló et al.8 treated 32 patients with 128 immediately loaded implants supporting fixed full-arch maxillary prostheses and obtained a
survival rate of 97.6% after 1 year. In a different study, the same authors treated 44 patients with 176 immediately loaded implants and fixed full-arch mandibular prostheses and obtained a survival rate of 97.1% after 1 year. Hinze et al. treated 37 patients with either mandibular or maxillary full-arch fixed prostheses supported by 4 implants, and at the 1-year follow-up the survival rates were 96.6% for maxillary implants and 98.7% for mandibular implants. Agliardi et al. published the largest series, with 61 maxilllas and 93 mandibles treated with fixed immediately loaded full-arch prostheses over 4 implants and followed for ≥1 year; 4 implants in the maxilla and 1 in the mandible failed, yielding respective survival rates at 1 year of 98.36% and 99.73%. In the present study, 6 patients were treated with mandibular and/or maxillary fixed full-arch prostheses over 4 implants; 20 implants were placed in the maxilla and 12 in the mandible. After an average follow-up of 22.9 months, the implant success rate was 100% according to the criteria of success described by Albrektsson et al. Radiologic criteria were evaluated in panoramic radiographs, because no intraoral radiographs could be taken owing to the extreme fragility of the oral mucosae.

Peñarrocha et al. compared patient satisfaction between fixed prostheses and overdentures over implants in patients with RDEB; satisfaction was high with both types of prosthesis, but slightly higher for fixed prostheses (9.6 vs. 8.8 with overdentures). In the present study, only fixed prostheses were used and average patient satisfaction was 9.0.

CONCLUSIONS

Fixed full-arch short-expand prostheses supported by 4 anterior implants can be successfully used to rehabilitate patients with RDEB, considerably improving these patients’ quality of life.

REFERENCES


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