Fixed Implant-Supported Full-Arch Prosthesis in Epidermolysis Bullosa With Severe Symptoms

Marcelo Guzmán Letelier, DDS
Claudia Crisosto Jara, DDS
Sonia Peñarrocha-Oltra, MD
Sonia Gomar-Vecher, DDS, PhD
Miguel Peñarrocha Diago, MD, PhD*

INTRODUCTION

Epidermolysis bullosa (EB) is a group of rare hereditary mucocutaneous disorders that is characterized by the presence of a mechanical defect between the skin and the mucosa membranes with recurring appearances of blisters and vesicles. This disorder is classified in 4 principal groups—Simplex, Junctional, Dystrophic, and Kindler syndrome—and 32 subtypes. Recessive dystrophic with generalized impact presents most frequently in the oral mucosa. Systemic features of this disease include blisters on the entire body—particularly in areas of friction, such as the hands, the feet, the elbows and the knees. The blisters subsequently burst, leaving painful ulcers that heal via contraction of the soft tissue. In its most severe presentations, junctional and dystrophic EB, this contraction of the soft tissue often results in digital syndactyly, with the formation of stumps and stenosis of the superior third of the esophagus; this, in turn, causes dysphagia. Oral symptoms include the formation of recurring blisters and scars, limited oral aperture, ankyloglossia, elimination of the mucosa, severe microstomia, ankyloglossia, and elimination of the mucosa. In some cases, rampant caries associated with hypoplastic enamel and poor oral hygiene are observed. The treatment of EB patients with conventional removable dentures has resulted in development of blisters in the oral mucosa as a result of mechanical friction of the prosthesis over the tissues. As a result, restoration using dental implants and a fixed prosthesis is likely to be better tolerated by the patient, as it limits the possibility of lacerations and blisters in the soft tissue.

The objective of this clinical case is to describe the restoration of the complete arch in an atrophic maxilla using endosseous implants and an implant-supported fixed prosthesis in a patient diagnosed with RDEB who is exhibiting severe general and oral symptoms. We will also evaluate improvements in the patient’s quality of life using a subjective scale of satisfaction.

PRESENTATION OF THE CASE

A 31-year-old woman, diagnosed with RDEB and severe symptoms, inquired about the possibility of restoration through endosteal implants in the maxilla. The patient had a history of stenosis of the esophagus with multiple and extensive scarring of the skin as a result of the contraction of bullae, digital syndactyly, cicatrical alopecia, and ectropion from the contraction of conjunctival lesions. The patient exhibited very low self-esteem and poor quality of life as a result of the disease, combined with the difficulty to eat due to stenosis and the soft diet required to present the formation of ulcers of the esophagus.

Intraoral clinical examinations revealed bleeding blisters in the mucosa, severe microstomia, ankyloglossia, elimination of buccal and vestibular sulci, advanced periodontal disease, and the lack of attached gingiva. In addition, multiple carious lesions were observed with notable deterioration of the dental tissue, significant destruction of dental crowns, apical lesions, and severe bone resorption with maxilla atrophy. Radiological exploration using computerized topography (CAT) revealed significant atrophy of the hard parts of the maxilla.

As the teeth were not recoverable, all remaining teeth in the maxilla were extracted with the exception of the second molars to preserve the vertical dimension (Figure 1a through c). Implant surgery was carried out under local anesthesia (articaine 4% and epinephrine 1:100 000) 3 months after the extraction, under conscious sedation with intraoral midazolam (5 mg/5 mL injectable solution). The lips and perioral tissue of the patient were constantly lubricated with liquid Vaseline. The implants were inserted using a low-velocity drill technique (without saline irrigation) combined with the use of osteotomes, given that conventional drilling could destroy the residual bone and reduce the retention of the implant. The
A low-velocity drill with no irrigation was used to create a small opening in the residual bone to allow insertion of osteotomes of small diameter. Whenever suction was required, the aspirator was placed in direct contact with the bone and not the mucosa, thereby avoiding formation of blisters. Six Zimmer TSV (Tapered Screw-Vent, Zimmer Biomet, Warsaw, Ind), 3.3, 3.7, and 4.7 in diameter were inserted in the bone.

**FIGURE.** Oral maxilla restoration using complete fixed prosthesis in a 31-year-old patient diagnosed with severe RDEB. (a) Facial aspect with multiple abrasions and scars on the skin as a result of blisters and the retraction of tissues. (b) Intraoral aspect: limited oral aperture, initial acrylic prosthesis in poor condition, displaced and lacking support. Multiple carious lesions with notable deterioration of the dental tissues making impossible the restoration over the natural tooth. (c) Initial CAT showing severe alveolar bone resorption with atrophy of the maxilla and considerable deterioration of the remaining teeth. Note the blocked right sinus.
anterior maxilla zone (from premolar to premolar). All inserts achieved primary stability greater than 35N/cm. Due to this satisfactory initial stability, the decision was made to leave the implants inserted into the oral cavity with healing caps. These were sutured using absorbable Vicryl 3.0 material (Figure 1d). The patient was prescribed oral antibiotics (amoxicillin + clavulanic acid 500–125 mg every 8 hours for 7 days) and nonsteroidal anti-inflammatory drugs (ibuprofen 600 mg every 8 hours for 4 days). Postoperative evolution was positive, with the wounds healing adequately. Both clinical and radiological investigation confirmed integration of the implants and that the peri-implant tissues were in good condition 6 months following the operation. Due to this positive evolution, oral restoration proceeded with the cementing of the fixed full-arch prosthetic over 6 endosseous implants.8,9,12,19

A small, customized acrylic tray was made based on the study model to take the dental impression using the closed tray technique with silicon for weight addition. Machined pillars were used to support a fixed total rehabilitation of cemented metal-ceramic structure (Figure 1e through h).

The patient was monitored after 1 month and every 3 months for a year following the insertion of implants and cementing of the prosthesis. Six months following the prosthetic restoration, the patient was asked to respond to a questionnaire to measure her satisfaction and the psychological impact of the state of her oral health. The variables measured were comfort and retention, function, appearance and aesthetics, phonation, and self-esteem. The questionnaire was explained in detail to the patient, who was to respond with a mark or point on the visual analogue scale (VAS) corresponding to her level of satisfaction or dissatisfaction. The VAS was calibrated with horizontal lines 10 cm in length, in which the left-hand side represented 0% (negative limit) and the right side 100% (positive limit). The follow-up period after the insertion of the implants was 12 months. During this time, there were no failures or loss of peri-implant bone.8,9,12,19

In April 2016, 3 years after the placement of maxillary implants, the mandible was treated. All remainder teeth were taken out, and 5 implants were placed (Figure 1i through k). The mandible has not been intervened since this year because the patient was able to eat without discomfort, allowing the aesthetics of her remainder teeth. However, due to the history of dental infections and progressive deterioration of the teeth, the mandible was also operated on in the same way as the maxilla in 2013.

In 2016, maxillary implants are in healthy conditions with no failure (Figure 1l and m).
DISCUSSION

EB is pathology with low worldwide prevalence. Fine and Hinter11 have suggested a prevalence of close to 45 patients per million inhabitants and an incidence close to 95 cases per million live births. As a result, there are very few published case studies regarding their treatment.

Furthermore, the pathology has a large range of forms and clinical manifestations. It is grouped into four principal categories—Simplex, Junctional, Dystrophic and Kindler syn-

**Figure continued.** (h) Facial aspect posttreatment. (i) Mandible intraoral aspect. Multiple carious lesions with notable deterioration of the dental tissues making impossible the restoration over the natural tooth. (j) Five implants inserted in the anterior region (between premolars) with good initial torque of over 40 N. (k) Panoramic radiograph after mandibular implant placement. Maxillary prosthesis can be observed. (l) Maxillary implants follow-up. Healthy mucosa after removing the prosthesis. (m) Facial aspect after mandible treatment.
drome—and 32 different subtypes.\textsuperscript{4,11} In RDEB, there are two subtypes: Hallopeau–Siemens and non-Hallopeau–Siemens.

The case presented (Hallopeau–Siemens) is a serious form with severe symptoms and results in a significant deterioration in the quality of life of patients, characterized by the development of blisters on the hands, feet, elbows, and knees; pseudo or digital syndactyly; stenosis of the esophagus; anemia; stunting; scarring alopecia; and increased risk of squamous cell carcinoma, principally in the hands and feet, combined with a greater impact of lesions in the oral mucosa and teeth deterioration.\textsuperscript{12–14}

We concur with the findings of Fine\textsuperscript{4} and Peñarrocha et al\textsuperscript{9} that the dental management of patients with severe RDEB represents a challenge for the oral surgeon, given that the oral mucosa exhibits blisters that heal through the formation of scars, causing microstomia, the obliteration of the buccal grooves, and ankyloglossia, all of which complicate surgical procedures and the use of prosthetics.\textsuperscript{12,15} Similar to these studies, we believe that delicate, nontraumatic treatment of the oral tissues and mucosa must be considered. In addition, lubrication of the lips and commissures with liquid Vaseline are recommended to reduce the risk of friction and tissue damage with the formation of blisters.\textsuperscript{16}

In this case, in accordance with reports by Wright\textsuperscript{17} and Peñarrocha and colleagues,\textsuperscript{12,15} surgical implant procedures must be carried out under local anesthesia and sedation, given that general anesthesia can increase the complexity due to ulceration caused by intubation (nonetheless, Lee et al\textsuperscript{3} have carried out surgery under general anesthesia). Local anesthesia injected deep into the mucosal tissues at a sufficiently slow velocity avoids distension, which can cause mechanical separation of the tissues and the formation of blisters.

In patients with EB, the literature highlights that when irrigating the operating field with sterile saline solution during the installation of implants, the contact between the aspirator and the oral mucosa can cause irritation of the tissues and the consequent formation of blisters.\textsuperscript{14,15} For this reason, in this case, the implants were inserted using the low-velocity drill technique without irrigation, dispensing with the need for cooling and, in turn, aspirating the saline; doing this did not result in bone loss during drilling.

In line with Peñarrocha and colleagues\textsuperscript{12,15} for patients with severe microstomia and considerable atrophy of the maxilla, implantation of implants in the anterior sector (canine buttress, plantine process, and residual alveolar bone) was carried out using osteotome technique, given that conventional perforation can destroy the residual bone process and reduce the retention of the implant. Only an initial drill with no irrigation is used to create a minimal aperture in the residual process to permit the insertion of osteotomes of small diameters.\textsuperscript{9,12,18}

The use of endosteal implants in the restoration of edentulous patients with RDEB could provide better results than traditional prostheses, given that the conventional removable dentures cause friction and irritation of the mucosa, which can lead to the development of blisters.\textsuperscript{9,12,19}

Dental restoration using a complete fixed and cemented prosthesis over 4–6 implants in edentulous patients with RDEB not only reduces the risk of trauma in the soft tissues by avoiding contact between the prosthesis and the oral mucosa, it also avoids the formation of blisters and reduces trauma to the esophagus by facilitating more efficient mastication. This improves the patients’ nutrition as they are able to adequately form boluses.\textsuperscript{12}

Given the limitation of buccal aperture, restoration with a fixed complete prosthesis offers the potential to manipulate the device with fewer incidences of trauma to the soft tissues, in addition to reducing the risk of involuntary swallowing prosthetic attachments, as screwdrivers are not required to assemble and disassemble the prosthesis. Furthermore, the distribution of the load is improved, tolerating greater structural passivity as it presents lower rates of mechanical complications when compared to screwed implants.\textsuperscript{20,21}

The literature describes 7 clinical series of patients with EB treated with dental implants.\textsuperscript{19} Seventeen patients underwent restoration with a total of 102 implants and a success rate of 97.7–100%.\textsuperscript{3,9,12,22} In this case, a comparable success rate of 100% was obtained. As EB is a disorder of the soft tissue and does not directly involve bone, it is reasonable that the success rates would be similar to those obtained in healthy patients. However, only case studies or clinical series with short follow-up periods have been published. As this case reports on only 1 clinical case, we understand the limitations of this study; however, we insist that it is possible to rehabilitate this type of patient successfully.

The visual analogue scale of satisfaction described earlier by Peñarrocha et al\textsuperscript{9} was used to evaluate the improvements in quality of life and self-esteem. For this, 5 variables were used: comfort, function, aesthetic, phonation, and self-esteem. Overall, this study averaged 9.5. Peñarrocha and colleagues compared the satisfaction of patients with RDEB between patients with implant-supported dentures and implant-supported fixed prosthetics, with values of 8.8 and 9.6, respectively. These results are fully consistent with the results that we obtained.

\section*{Conclusion}

This treatment option appears to be favorable for patients with RDEB with severe symptoms when compared to alternatives that involve removable dentures that irritate the oral mucosa.

\section*{Abbreviations}

EB: epidermolysis bullosa
RDEB: recessive dystrophic epidermolysis bullosa
VAS: visual analogue scale

\section*{References}