

Restoration with Implants in Patients with Recessive Dystrophic Epidermolysis Bullosa and Patient Satisfaction with the Implant-Supported Superstructure

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Purpose: The use of endosseous implants in the prosthetic restoration of edentulous patients with recessive dystrophic epidermolysis bullosa (RDEB) may provide improved outcomes when compared with traditional prosthetic methods. The aim of this study was to evaluate the feasibility of placing endosseous implants in patients with RDEB and to compare the treatment outcomes of fixed and removable implant-supported restorations in the edentulous maxilla or mandible with the main emphasis on patient response. **Materials and Methods:** Six patients with RDEB were treated with implants. All patients were completely edentulous in either the maxilla or mandible and had marked oral involvement, with alterations in the soft and hard tissues in all cases. Three patients were treated with fixed, screw-retained implant-supported prostheses, and 3 were treated with removable implant-supported prostheses. Six months after prosthetic restoration, patients were given a questionnaire to assess their psychologic well-being and satisfaction with the implant-supported restoration marked on a visual analog scale. **Results:** A total of 38 dental implants (21 maxillary, 17 mandibular) were placed in 6 patients. The implant success rate was 97.9%. The average follow-up from implant placement was 5.5 years (range, 1 to 9). The fixed and removable implant-supported prostheses were associated with improvements in comfort and retention, function, esthetics and appearance, taste, speech, and self-esteem. The level of satisfaction was slightly higher in patients with a fixed prosthesis. **Conclusion:** These findings suggest that endosseous implants can be successfully placed and provide support for prostheses in patients with RDEB. Patients with fixed prostheses and overdentures were satisfied with their implant-supported prostheses in the edentulous maxilla and mandible. (Case Series) INT J ORAL MAXILLOFAC IMPLANTS 2007;22:651-655

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Epidermolysis bullosa (EB) is a group of mainly hereditary skin disorders manifested by a ten-

density of the skin and mucosa to form bullae and vesicles following minor friction and trauma. The disorder is classified into 3 major presentations (simplex, junctional, and dystrophic) and 25 subtypes—the recessive dystrophic form of EB with generalized involvement (RDEBg or the Hallopeau-Siemens variant) being the presentation with the greatest oral mucosal involvement.¹

Oral features include repeated blistering and scar formation leading to limited oral opening, ankyloglossia, elimination of buccal and vestibular sulci, perioral stricture, severe periodontal disease, alveolar bone resorption, atrophy of the maxilla with mandibular prognathism, an increased mandibular angle, and predisposition to oral carcinoma. Routine dental care or even normal tooth brushing might cause bullae on the oral mucosa.²

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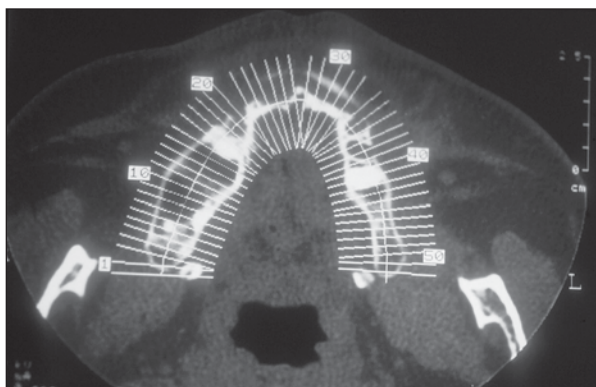


Fig 1 Case 3: Extensive bone atrophy in the maxilla.

Systemic features include bullae formation on hands, feet, elbows, and knees. The lesions are initially noted at or soon after birth. Bullae leave painful ulcers on rupturing, and healing is often followed by scarring and tissue contraction. Contractures and syndactyly of digits might result in the formation of claw-like hands. The upper esophagus frequently becomes stenotic, leading to dysphagia or esophageal obstruction.^{3,4}

The aim of this study was to determine whether endosseous implants can be successfully placed, restored, and loaded in patients with recessive dystrophic EB (RDEB) and to determine the degree of patient acceptance and satisfaction.

MATERIALS AND METHODS

Forty-two patients with RDEB presenting for dental treatment were examined between 1995 and 2004. The inclusion criteria were a diagnosis of RDEB, an association with total edentulism involving at least the maxilla or mandible, the acceptance of implant treatment, and a minimum follow-up of 1 year after implant loading. Radiographic records consisted of panoramic film and computerized tomographic (CT) scans of the jaws, which revealed extensive bone atrophy (Fig 1).

Surgery was performed with the patient under local anesthesia and conscious sedation. The lips were lubricated with petroleum jelly to avoid tissue friction or irritation of the mucosa and bulla formation. When intraoral local anesthesia was administered, the anesthetic solution was injected deeply into the tissues at a rate slow enough to prevent tissue distortion, which may cause mechanical tissue separation and blistering. The placement of maxillary implants required the use of an expansion osteotome because conventional mechanized instruments would have destroyed the entire residual bony

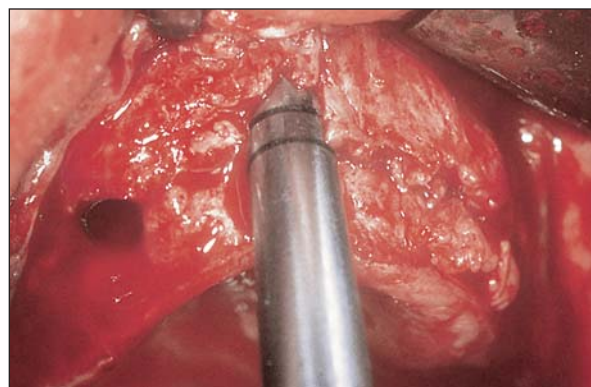


Fig 2 Case 3: The creation of the implant bed with the osteotome.

process, thus precluding primary retention of the implants (Fig 2). In contrast, in the mandible, drills and irrigation were required to prepare the implant beds. Both Straumann implants (Institut Straumann, Basel, Switzerland) and Centerpulse implants (Centerpulse Dental, Carlsbad, CA) were placed.

Oral antibiotics (amoxicillin, 500 mg every 8 hours for 7 days) and nonsteroidal anti-inflammatory medication (ibuprofen, 600 mg every 8 hours for 3 days) were administered. The implants were allowed to integrate for a period of 3 months in the mandible and 6 months in the maxilla before prosthetic loading. Patients with 2 or 3 implants were restored with overdenture-type prostheses, and patients with 4 or 5 implants were restored with implant-supported fixed prostheses. The patients were clinically monitored after 1 month, after 3 months, and every 6 months thereafter.

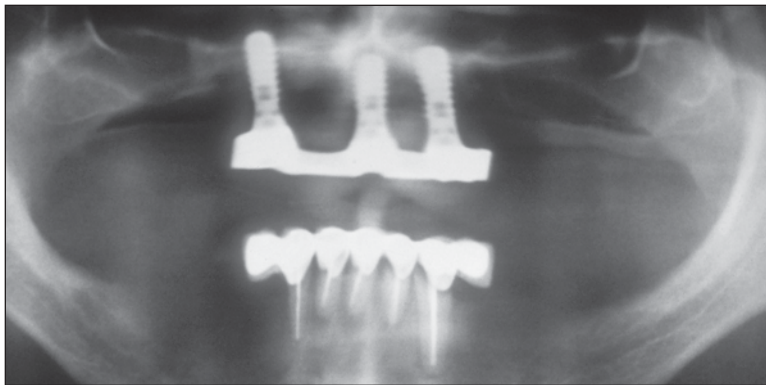
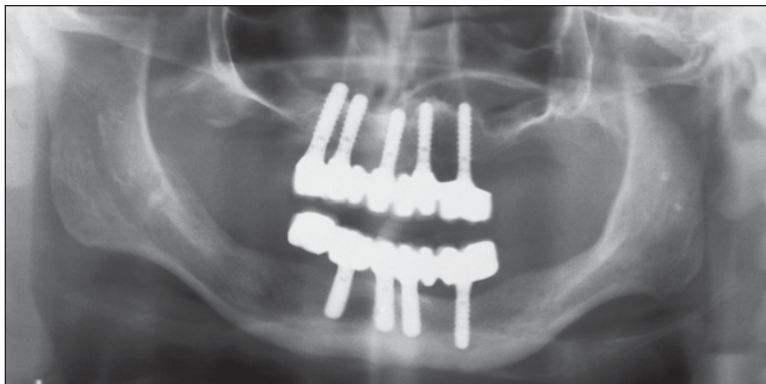
Six months after prosthetic restoration, patients were asked to complete a questionnaire measuring their satisfaction and the psychological impact of their oral health status. The parameters chosen were comfort and retention, function, esthetics and appearance, taste, speech, and self-esteem. The questionnaire was carefully explained to the patient, and he or she was asked to place a mark at a point on a visual analog scale (VAS) corresponding to his or her level of 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).

RESULTS

Of 42 patients with RDEB treated within the study period, 6 patients (4 women and 2 men, aged 23 to 44 years) underwent prosthetic restoration involving implant-supported prostheses. All patients exhibited marked oral involvement, with devastating alter-

Table 1 Clinical Data of the Patients with RDEB

Case	Age	Sex	Edentulism	No. of implants			Failures	Overdenture		Fixed prosthesis		Follow-up (y)
				Mx	Mnd	Total		Mx	Mnd	Mx	Mnd	
1	23	F	Mnd	-	2	2	0		X			9
2	36	F	Mx, Mnd	4	2	6	0	X	X			8
3	28	M	Mx	3	-	3	0	X				7
4	44	F	Mx, Mnd	5	4	9	0			X	X	5
5	29	M	Mx, Mnd	5	4	9	0			X	X	3
6	43	F	Mx, Mnd	5	4	9	1 Mx			X	X	1

**Fig 3** Case 3: Restoration with a bar-retained overdenture.**Fig 4** Case 3: Prosthetic restoration of an edentulous patient with an overdenture prosthesis. Note formation of bleeding bullae in tongue and in lips.**Fig 5** Case 5: Restoration with a fixed prosthesis.**Fig 6** Case 5: Oral features, repeated blistering, and scar formation led to limited oral opening.

ations in the soft and hard tissues in all cases, and all had antecedents of bleeding blisters, dental caries, and loss of dentition. Microstomia, assessed using the classification of Naylor et al,⁵ was severe in all cases, as was obliteration of the oral vestibule and ankyloglossia.

A total of 38 implants were placed, 29 Straumann implants and 9 Centerpulse implants. Twenty-one implants were placed in the maxilla with the osteotome technique; the alveolar ridges were atrophic in all cases. Seventeen implants were placed

in the mandible using conventional osteotomy preparation (Table 1).

Surgical management was complicated by bleeding bullae as a result of minor trauma. Blister complications were recorded during the operation in all cases, especially in relation to implants positioned in the mandible, although the postoperative course was uncomplicated. Three patients were provided with overdentures (cases 1, 2, and 3; Figs 3 and 4), and 3 were provided with complete fixed prostheses (cases 4, 5, and 6; Figs 5 and 6).

Table 2 Mean VAS Ratings of Various Aspects of Treatment 6 Months After Prosthesis Delivery

Case	Comfort	Function	Esthetics	Taste	Speech	Self-esteem	Total mean
Overdentures							
1	10	9	9	8	10	9	
2	9	7	8	8	7	8	
3	9	9	9	10	10	9	
Mean	9.3	8.3	8.7	8.7	9.0	8.7	8.8
Fixed prostheses							
4	10	9	10	10	9	6	
5	10	10	10	10	10	10	
6	10	10	10	10	10	10	
Mean	10.0	9.6	10.0	10.0	9.6	8.7	9.6

One implant failed to achieve osseointegration. At successive follow-ups, oral mucosal ulcerations were frequently observed in areas in contact with the overdentures. However, the peri-implant mucous soft tissues remained in good condition in all patients. Radiographs were obtained every 12 months, and the radiologic follow-up confirmed osseointegration of the implants; no untoward peri-implant bone loss was recorded. The average follow-up for patients was 5.5 years (range, 1 to 9), and the success rate was 97.9%.

Before prosthesis placement, the patients were unable to chew, and all food had to be swallowed in purée form to avoid esophageal ulceration. After prosthetic restoration, all patients were able to chew and swallow a well-ground food bolus. Patient quality of life improved considerably as a result of treatment, esthetically, socially, and functionally, by allowing mastication (Table 2). Both prosthetic designs were associated with significant improvements in comfort and retention, function, esthetics and appearance, taste, speech, and self-esteem. With respect to patient assessment of implant therapy, the level of satisfaction was very high in both groups. It was slightly higher in the fixed prosthesis group (mean 9.6) than in the overdenture group (mean 8.8).

DISCUSSION

In patients with the most severe form of RDEB, the oral mucosa exhibits marked blistering that heals with scar formation, causing microstomia, obliteration of the oral vestibule, and ankyloglossia. Totally edentulous patients may benefit from restoration with implant-supported prostheses because conventional complete prostheses may cause tissue irritation and bulla formation secondary to friction from complete dentures that would not, under normal circumstances, be pathogenic.

All of patients in this study were treated using local anesthesia and sedation because general anesthesia poses the added problem of possible ulcerations resulting from intubation.^{3,6} Surgery for patients with RDEB is complicated by the formation of bleeding bullae as a result of minor trauma. Implant surgery poses an additional problem because flap elevation can produce bullae. Copious irrigation with sterile saline is recommended, but aspiration of this fluid may cause contact between the aspirator and oral mucosa, inducing considerable tissue irritation with the formation of bullae. For hard tissue management, all 6 patients presented significant bone atrophy. In the maxilla, osteotomes were used to allow expansion of the alveolar process with primary fixation in crests with marked alveolar atrophy. In the mandible, the conventional rotary procedure was used. Care was taken to secure a surgical field of sufficient size to allow work on the bone without inducing soft-tissue tension. Aspiration was performed with the aspirator contacting bone rather than the soft tissues.

Both prosthetic designs were associated with improvements in patient assessment of comfort, retention, function, esthetics and appearance, taste, speech, and self-esteem. Zitzmann et al treated patients with fixed implant-supported prostheses in the maxilla or mandible, and both groups were associated with significant improvements in comfort and retention, function, esthetics and appearance, taste, speech, and self-esteem.⁷

Epidermolysis bullosa severely affects quality of life in patients with this disease.⁸ The use of endosseous implants in the prosthetic restoration of edentulous patients with RDEB may provide a considerably improved outcome compared with traditional methods.^{9,10} The 6 patients described in this study related improvements from the esthetic and social perspective; more importantly, however, the new teeth allowed them to chew and swallow food

without the formation of esophageal ulcers. The results of this study suggest that implants can be successfully placed for subsequent prosthetic restoration in these patients. Patients indicated a high level of satisfaction, which was slightly higher in those with fixed dentures than those with overdentures.

CONCLUSION

The results of this retrospective review of the clinical performance of 6 patients suggest that endosseous implants can be successfully placed and used to support dental prostheses in patients with RDEB. Patients described satisfaction with both fixed and removable prostheses.

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