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Oral Rehabilitation of a Patient with Papillon Lefèvre Syndrome Using Fixed Full-Arch Hybrid Prostheses Supported by Four Axially Loaded Implants: A Case Report with Four-Year Follow-up

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A B S T R A C T

This report details the successful prosthetic rehabilitation of a 25-year-old male patient with Papillon Lefèvre Syndrome (PLS) using an implant-supported hybrid prosthesis. Six implants were placed in the maxilla, and four were placed in the mandibular arch. All implants were inserted axially (non-tilted) and were planned to be loaded after a healing period of 6 months. One implant failed due to graft loss during the healing phase, which was removed and the remaining implants were restored with a hybrid prosthesis after 6 months, using the delayed loading protocol. The patient was followed-up for four years and all the remaining implants successfully integrated and remained fully functional during this period. The prosthesis significantly improved the functional, aesthetic, and psychological well-being of the patient. This case report is the first of its kind to use only four axially placed implants for rehabilitation of a PLS patient with a successful four-year follow-up.

Keywords: Dental Implants; Dental Prosthesis, Implant-Supported; Mouth Rehabilitation; Papillon-Lefevre Disease

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INTRODUCTION

Papillon–Lefèvre syndrome (PLS) is a rare autosomal recessive disorder, which presents with palmar-plantar hyperkeratosis, or thickening of the soles and palms, and severe destructive periodontal disease affecting both deciduous and permanent teeth. The etiology and pathogenesis of the disease are still not clear, but patients with PLS were found to have mutations of the Cathepsin C gene localized in chromosome 11q14 [1]. Cathepsin C is highly expressed in keratinized epithelial tissues such as the

palms, soles, and gingiva, as well as in defense cells like neutrophils, T lymphocytes, and natural killer cells. It plays a critical role in both epithelial differentiation and immune activity. Inactivation of Cathepsin C due to mutation causes a reduction of enzymatic activity resulting in an array of clinical symptoms associated with PLS [1]. Decreased epithelial differentiation leads to hyper-keratosis of palms and soles. Periodontal breakdown and early tooth loss occur due to reduced response of neutrophils to combat oral infection by *Staphylococcus* spp. and *Aggregatibacter actinomycetemcomitans* [2]. PLS patients are also more prone to liver/renal abscess, sinusitis, and respiratory tract/urinary infections due to decreased immune response [2].

Prosthetic rehabilitation of PLS patients with removable dentures [3], overdentures, and Cu-Sil dentures [4] have been tried in the past. Implant-supported prostheses have been shown to be successful in rehabilitation of patients with PLS with improved function and good long-term prognosis [5]. Rehabilitation of PLS patients with multiple implant-supported fixed full-arch prosthesis has been previously reported [6]. Due to severe bone resorption in PLS patients, placement of dental implants is often challenging and may require bone augmentation procedures. Kinaia et al. [7] used calvarium bone graft for augmenting severely atrophied ridges in PLS patients followed by endosseous implant placement. Use of short dental implants followed by implant-supported removable prosthesis has been tried as an alternative strategy to manage severely atrophied mandibular ridges in PLS patients [8].

This case report presents a rare case of prosthetic rehabilitation of a 25-year-old edentulous male patient with PLS using only four axially placed implants in the anterior mandible and six implants in the maxilla. Prosthetic rehabilitation was done using fixed implant-supported hybrid denture in both arches.

CASE REPORT

A 25-year-old male patient with PLS was referred from the Department of Oral and Maxillofacial Surgery to the Department of Prosthodontics and Implantology after he had undergone a maxillary ridge augmentation surgery using iliac crest graft. Vertical and horizontal maxillary ridge augmentation had been done before nine months. After the healing period, he was referred to our department for the prosthetic phase. His chief complaint was difficulty in chewing food, lack of aesthetics, and multiple missing teeth in both arches since childhood. According to the patient's dental history, he had been wearing removable dentures since childhood, but expressed dissatisfaction with them. This patient experienced emotional and psychological distress as a result of his early tooth loss and resulting disability.

Examination revealed diminished lower facial height, mid-facial deficiency, pseudo-class III jaw relationship, and hyperkeratosis of the palms and feet. Intraoral findings consisted of a completely edentulous maxilla and partially edentulous mandibular arch with all teeth missing except #17 and #32 (Fig 1).

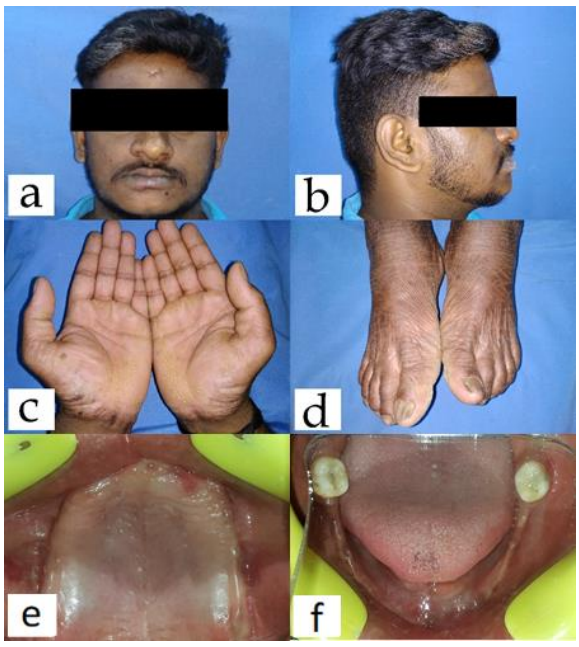

Fig 1. Clinical findings of the Papillon-Lefevre patient. a) pre-operative frontal extra-oral view, b) pre-operative extra-oral view of the profile, c) hyperkeratosis of the palms, d) hyperkeratosis of the feet, e) pre-operative intra-oral view of the maxilla, f) pre-operative intra-oral view of the mandible

Pre-operative orthopantomogram (OPG) showed maxillary augmentation with graft stabilizing pins. The maxillary arch had sufficient width after the augmentation procedure while the mandibular arch was atrophic with insufficient bone height in the posterior mandible (Fig 2).

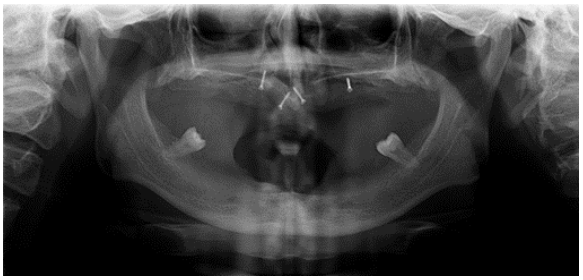

Fig 2. Pre-operative orthopantomogram showing maxillary augmentation with graft stabilizing pins and right and left mandibular third molars in an atrophic arch

The remaining teeth (#17 and #32) in the mandible had pocket depths of 3-4 mm with no mobility. The mucosa was smooth, firm, and keratinized with absence of stippling, and salivary flow was normal. The patient did not have any other systemic disease and familial history did not indicate similar symptoms in any of the patient's family members.

In the initial phase, we planned to fabricate conventional complete dentures to evaluate the aesthetics and ridge relations. Conventional dentures were fabricated at the appropriate vertical dimension and the patient's approval for aesthetics was obtained. Various treatment options like continuation of removable complete dentures, implant over dentures, and implant supported hybrid dentures were given to the patient. He opted for implant-supported hybrid prosthesis but did not want to extract his mandibular third molars. Hence, they were left untouched and were planned to be left out of occlusion from the final prosthesis. The risks, benefits, duration, and cost of the procedure were clearly explained and his consent was obtained.

Prior to surgery, oral prophylaxis was performed, and the patient was prescribed an oral rinse of 0.2% chlorhexidine for two weeks. A full-thickness flap was reflected and six implants (Touareg–S; Adin Dental Implant System Ltd, Israel) were placed in the maxilla. All implants were placed axially (non-tilted) using a freehand technique, without the use of surgical guides. In the maxillary right quadrant, a total of three implants measuring 4.2mm×11.5mm were placed. In the maxillary left quadrant, implants measuring 4.2mm×10 mm, 4.2mm×11.5 mm, and 5mm×10 mm were inserted starting from the midline. After implant placement, healing abutments were connected and sutured (Fig 3a, b). A full-thickness flap was elevated in the mandibular anterior region (Fig 3c) and crestal osteotomy of 3-4 mm was performed to attain sufficient width (Fig 3d). Four 4.2mm×10mm implants (Touareg–S; ADIN Dental implant system Ltd, Israel) were placed between the two mental foramina regions (Fig 3d). All implants were placed axially (non-tilted) and no surgical guides were used. Cover screws were placed and primary closure was achieved.

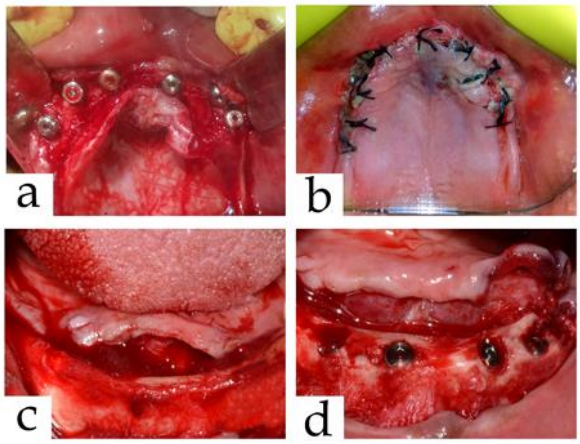

Fig 3. Surgical procedure and implant placement. a) maxillary implants connected to healing abutments, b) primary closure with sutures in the maxilla, c) thin alveolar crest in the mandibular anterior region, d) implants placed in mandibular anterior region after crestal osteotomy

To avoid any potential implant failure, the prosthetic phase was scheduled six months after the initial healing period, due to the patient's PLS condition. At the sixth month visit, the patient complained of pain in the left maxillary anterior region. OPG taken during the review showed a radiolucency around the apex of the implant in tooth #10 region (Fig 4) suggesting implant failure. Clinical examination revealed implant mobility and hence it was removed under local

anesthesia. The implant was not replaced, as there was graft loss and the patient did not agree to another grafting procedure in the same region. Mandibular implants were exposed in a second stage surgery and healing abutments were connected.

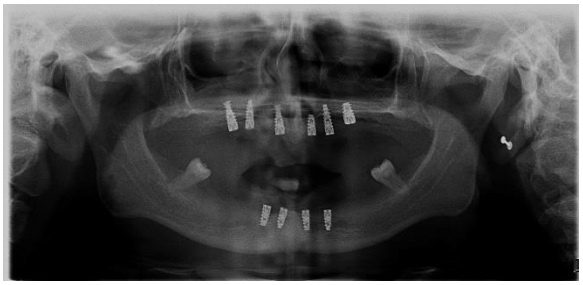

Fig 4. Follow-up orthopantomograms taken six months after implant placement

Preliminary impressions were made using irreversible hydrocolloid material (Zelgan Plus; Dentsply Sirona) and casts were poured. Custom open trays were fabricated with auto polymerizing polymethyl methacrylate resin (DPI Cold Cure; Dental Products of India-RR) and open tray implant impressions were made with heavy body and light body elastomeric impression material (Aquasil Heavy Body, Light body; Dentsply Sirona). After the final casts were obtained, a verification jig was fabricated and the fit was checked intraorally and radiographically.
The position of the metal framework was determined using tentative jaw relation records and trial dentures. Briefly, light cure denture bases were fabricated on the casts and occlusal rims were attached to the denture bases (Fig 5a). Occlusal rims were adjusted to optimal vertical dimension and aesthetics. A tentative jaw relation record was obtained and the casts were mounted on a semi-adjustable articulator (Fig 5b). Trial dentures were fabricated and tried in the patient (Fig 5c). The space and position of the bar was evaluated using these trial dentures (Fig 5d).
Metal frameworks made of cobalt-chromium alloy (Wironium alloy, Bego, Germany) were then fabricated and tried in the patient (Fig 5 e, f). Bite rims were attached to the frameworks and the optimal vertical dimension was re-established. Incisal visibility, freeway space, speaking space, buccal corridor space, and phonetic evaluations were done at the established vertical dimension. The patient was guided into the centric relation using the bimanual manipulation method and bite registration was made using polyvinyl siloxane bite registration material (Jet Bite, Coltene/Whaledent, Switzerland).

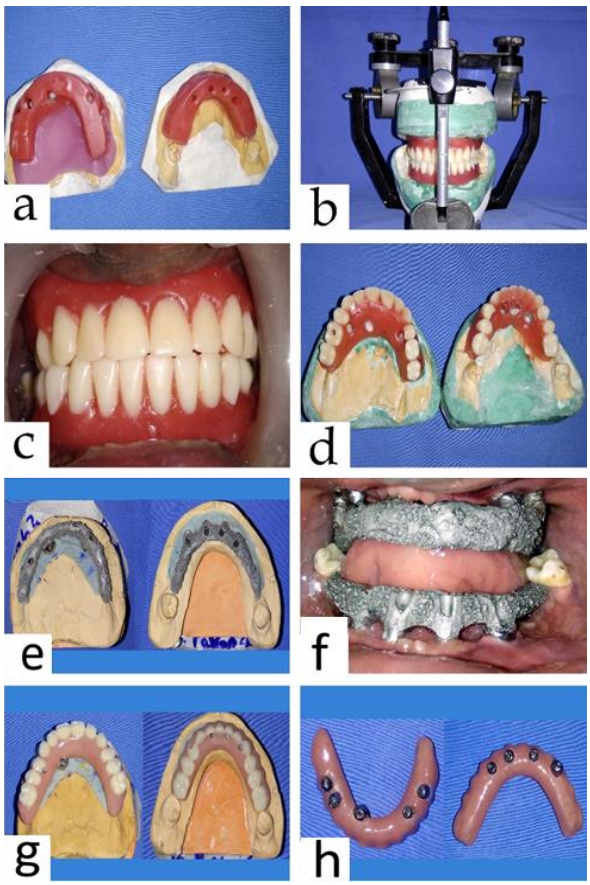

Fig 5. Laboratory procedures for prosthetic rehabilitation. a) temporary denture base with occlusal rims attached, b) casts mounted on semi-adjustable articulator with trial dentures, c) try-in of dentures in the patient, d) evaluation of space for the bar using trial dentures, e) maxillary and mandibular Co-Cr frameworks, f) framework trial, g) occlusal view of the prosthesis, h) tissue surfaces of the prosthesis

Maxillary and mandibular casts were mounted on a semi-adjustable articulator (Hanau Wide-Vue Arcon Articulator, Whip Mix Corporation, Louisville, USA) in centric relation using the bite record. Semi-anatomic teeth (Acry Rock; Ruthinium Dental Products Pvt. Ltd., New Delhi, India) were arranged and extended only till the first molar region in both maxilla and mandible to limit the cantilever length (Fig 5 c, d). Trial

denture was checked in the patient and his approval was sought for aesthetics and phonetics. After acrylization, finishing and polishing, the final prosthesis was inserted with a tightening torque of 15Ncm using a torque wrench (Fig 6 a-c). Care was taken to maintain a minimum of 2-3mm space between the hybrid prosthesis and the tissues to facilitate oral hygiene procedures.

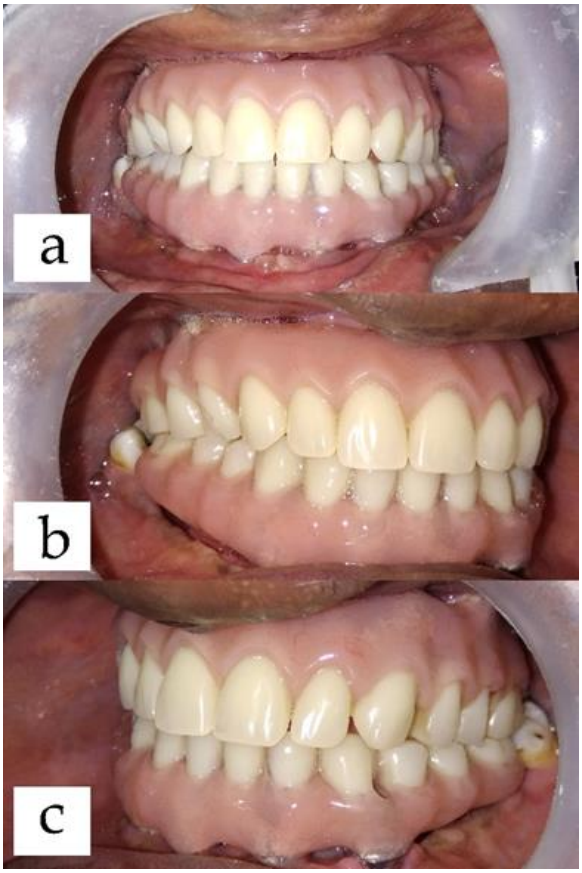

Fig 6. Post-operative intra-oral view of the hybrid prosthesis a) frontal view, b) right lateral view, c) left lateral view

Maintenance and follow-up:

The patient was instructed to come for at least one annual check-up and was taught to maintain oral hygiene using a water flosser. He was followed-up for a period of four years after implant placement and was comfortable with the hybrid prosthesis and was satisfied with its phonetics, retention, stability, and aesthetics. During the fourth-year follow-up, the patient reported no pain or discomfort, oral hygiene, and denture maintenance were satisfactory. No implants were mobile or lost and the follow-up OPG also showed only minimal bone loss around the implants (Fig 7).

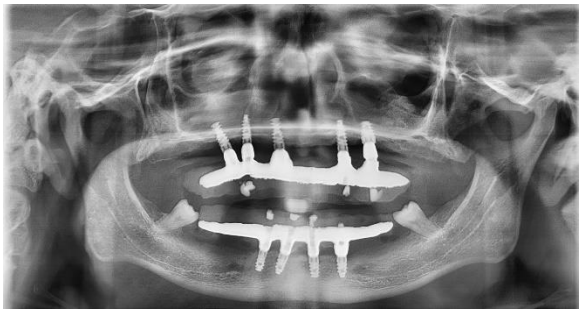

Fig. 7. Follow-up orthopantomograms taken four years after prosthetic rehabilitation

DISCUSSION

This report presents a unique case of a 25-year-old male patient with PLS who underwent prosthetic rehabilitation with implant-supported hybrid prosthesis, using only four axially placed implants, and was followed-up for four years. It is the first of its kind to document such a case. This is in contrast to the concept of using a combination of two tilted and two axially placed implants (all-on-four concept) as described by Maló et al. [9].

There are very few case reports of using only four axially placed (non-tilted) implants for full arch rehabilitation with a delayed loading protocol. This protocol has been tried previously for patients with Epidermolysis Bullosa (EB) who had fragile mucous membranes [10-13]. Penarrocha et al. [10] evaluated the feasibility of rehabilitating EB patients using endosseous implants and a delayed loading protocol. They found that out of the 27 placed implants (15 maxillary and 12 mandibular) only one failed and the rest integrated successfully during an average follow-up of 3 years [10]. Other clinical reports by Lee et al. [11], Muller et al. [12] and one study by Peñarrocha-Oltra et al. [13] have also indicated good success rates using this protocol.

Rehabilitation of PLS patients with dental implants is challenging due to unpredictable clinical outcomes and limited availability of bone. In our case we have used only 4 implants in the anterior mandible and 6 implants in the maxilla,

out of which one implant in the maxillary arch failed prior to loading after 6 months. Two recent systematic reviews reported similar success rates of close to 84% for implant-supported prosthesis in PLS patients [5,14]. Atarbashi-Moghadam et al. [14] reported that the failure rate of implants in the upper jaw (9%) was more than the mandible (2%) in PLS patients. A delayed loading protocol was adopted to prevent complications and implant failure. Nassani et al. [5] in their systematic review on survival rates of dental implants in individuals with PLS reported that out of the 10 included studies, nine of them followed only the delayed loading protocol. Although the success rates of implants in PLS patients are lower compared to those without the disease, most previous studies have reported that it can still be a viable treatment option with a good prognosis. [3,5-7,14].

CONCLUSION

Our patient with PLS was successfully rehabilitated with full-arch hybrid prosthesis supported by four axially loaded implants in the mandible and six implants in maxilla. Only one implant failed in the maxillary arch and all the remaining implants were fully functional during the four-year follow-up period. The prosthesis significantly improved the functional, aesthetic, and psychological well-being of the patient.

CONFLICT OF INTEREST STATEMENT

None declared.

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