Full-Mouth Rehabilitation with Calvarium Bone Grafts and Dental Implants for a Papillon-Lefèvre Syndrome Patient: Case Report

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Papillon-Lefèvre syndrome (PLS) is a rare autosomal recessive disorder of keratinization associated with palmoplantar keratoderma and severe periodontitis resulting in complete edentulism in late adolescence. The pathognomonic dental features of PLS are pathologic migration, hypermobility, and exfoliation of the teeth without any signs of root resorption. It has been suggested that an effective way to treat PLS patients presenting early in the disease progression is extraction of the erupted primary dentition or hopeless permanent teeth followed by antibiotic coverage with periodontal therapy for the remaining teeth. Unfortunately, studies have shown that this regimen only temporarily delays the progression of periodontal disease and does not prevent further tooth loss and bone destruction in the long term. Post–tooth loss, atrophic ridges make conventional prosthodontic rehabilitation quite challenging, and more recently, implant-supported prostheses have been considered as a viable alternative. In a PLS patient, implant placement is complicated by inadequate bone volume; thus, bone augmentation techniques or the use of short implants is often considered. When large volumes of bone are required, parietal calvarium bone can be used to predictably reconstruct severe defects. A PLS patient aged 21 years presented a chief complaint of ill-fitting conventional complete dentures. The patient had severely atrophic ridges, requiring significant bone augmentation for an implant-supported prosthesis. The present case is the first example of bone augmentation using autogenous calvarium parietal graft followed by endosseous implant placement and prosthetic restoration in a PLS patient. Int J Oral Maxillofac Implants 2017;32:e259–e264. doi: 10.11607/jomi.6282

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Papillon-Lefèvre syndrome (PLS) is a rare autosomal recessive disorder of keratinization.[1] Excessive keratin deposition is observed most prominently on the palms of the hands and soles of the feet, also known as palmoplantar keratoderma. Genetic studies have revealed that the loss of cathepsin-C function due to a gene mutation in chromosome 11q14.1 is strongly associated with PLS.[2] With a prevalence of 1 to 4 cases per million and no known sex or race predilection, this condition does not present routinely in periodontal practice; however, when it does, patients display significant oral changes. Consanguinity of parents is associated with PLS incidence; however, there is rarely a reported family history of disease.[3] Upon eruption of the primary dentition, the gingiva becomes inflamed, and a rapid destruction of the periodontium follows, resulting in premature loss of the deciduous teeth. The same process occurs during eruption of the permanent dentition, resulting in complete edentulism in adolescence.[4,5] The pathognomonic dental features of PLS are pathologic migration, hypermobility, and exfoliation of the teeth without any signs of root resorption.[4,5]

No difference has been reported in the oral bacterial flora of PLS-affected individuals compared with chronic periodontitis patients, with gram-negative cocci, rods, and spirochetes being common.[6] Neutrophil phagocytosis, impaired bactericidal activity, and decreased cell migration have been suggested as factors involved in the increased susceptibility to periodontal disease and infection in PLS cases.[7] The rapid

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periodontal destruction is not arrested by traditional periodontal therapy alone, leading to significant bone loss and severely atrophic arches.[8] The literature cites different management protocols for patients with PLS. Tinanoff et al reported effective therapy of PLS in a primary dentition patient with recommendation of extractions of the erupted primary teeth and antibiotic coverage. The 6- and 15-year follow-up showed no effect of PLS on the permanent dentition, with no detected polymorphonuclear leukocyte (PMN) dysfunction.[9,10] With regard to the PLS patient having permanent dentition, extractions of hopeless teeth combined with antibiotic coverage, periodontal therapy, and effective oral hygiene can slow the progression of periodontitis, but this is only temporary, resulting in eventual tooth loss.[11]

Atrophic ridges make conventional prosthodontic rehabilitation quite challenging, and more recently, implant-supported prostheses have been considered as a viable alternative.[12] Implants can improve the support, stability, and retention of prostheses, but to date, there have been very few cases available in the literature with successful application of implant therapy in PLS patients.[1,12–15] In a PLS patient, implant placement is complicated by inadequate bone volume. These patients generally have complete alveolar bone loss with only basal bone present. Thus, extensive bone augmentation using extraoral sites is often employed. Extraoral sites may include tibia, ribs, iliac crest, or calvarium bone.[16] Bone harvested from the calvarium contains thick cortical bone with minimal cancellous bone, resembling the bone characteristics of the maxillofacial complex. The rigidity of the cortical bone creates excellent space maintenance to reconstruct the deficient alveolar bone and can be supplemented with additional cancellous bone to fill all dead spaces.[16] Therefore, when large volumes of bone are required, autogenous calvarium bone can be a predictable source to reconstruct severe defects.[16] Although an extraoral surgical site and general anesthesia are required with calvarium grafting, major complications at the donor site are rare.[17] The thickness of the cranial bones varies with age, exhibiting a thick pericranium until age 3 years, becoming thinner and almost disappearing after age 40 years. Tessier reported in a case series that some patients had extremely thin bone, while others had over 12 mm bone thickness, and up to 32 mm in one case.[16] The cranial bone thickness requires preoperative assessment through three-dimensional radiographic evaluation. The present case shows the first example of successful bone augmentation with the use of autogenous calvarium parietal graft followed by endosseous implant placement and prosthetic restoration in a male PLS patient aged 21 years completed in faculty and private practice settings.

MATERIALS AND METHODS

Clinical Presentation

A male patient aged 21 years presented to the periodontal clinic at the Boston University Institute for Dental Research and Education (BUIDRE) with complete edentulism. The patient presented with a concave facial profile and complained of inability to chew food properly and was interested in a dental implant–supported prosthesis (Figs 1a and 1b). Medical history revealed Papillon-Lefèvre Syndrome with an otherwise unremarkable medical record. The patient exhibited palmar and plantar hyperkeratosis on the extremities and reported a difficult lifestyle (Figs 1c and 1d). Dental history revealed complete loss of teeth at age 13 years, with three sets of complete maxillary and mandibular dentures over the years. Clinical examination revealed extremely atrophic ridges (Fig 1e). Radiographically, cone beam computed tomography (CBCT) showed primarily basal bone that was inadequate for dental implant placement (Fig 1f). The patient was referred to the oral surgeons at French Dental Clinic for cranium parietal bone graft technique evaluation.[18] Further radiographic evaluation using a medical computed tomography (CT) scan (Fig 2a) was conducted, revealing adequate bone skull density of the cortical external and internal surfaces for cranium graft to reconstruct the atrophic maxilla and the mandible. Complete rehabilitation of the maxilla and mandible using a cranium graft for a dental implant–supported prosthesis was recommended, and the patient consented to treatment.

Cranium Parietal Bone Graft Surgery

The cranium parietal bone graft surgery was performed under general anesthesia with A.Z. and J.F.T. The maxillary and mandibular recipient sites were prepared, including bilateral maxillary sinus elevation procedures. The patient cranium (donor site) had adequate bone thickness for multiple cortical parietal bone grafts from the external surface (Fig 2b). The cranium mimics the bone characteristics in the maxillofacial area, and therefore, possesses similar resorption rates compared with intraoral grafts. In addition, grafts harvested from the cranium result in less morbidity such as pain, difficulty in walking, or potential body deformity compared with grafts taken from the tibia or iliac crest.[16] The parietal bone grafts were secured to the maxillary and mandibular arches using surgical fixation screws (Fig 2c). Complete primary closure was obtained for recipient and donor sites, and the patient was followed up in the hospital for 2 weeks before being released (Fig 2d). The area was allowed to heal for an additional 3 months prior to reentry for implant placement.

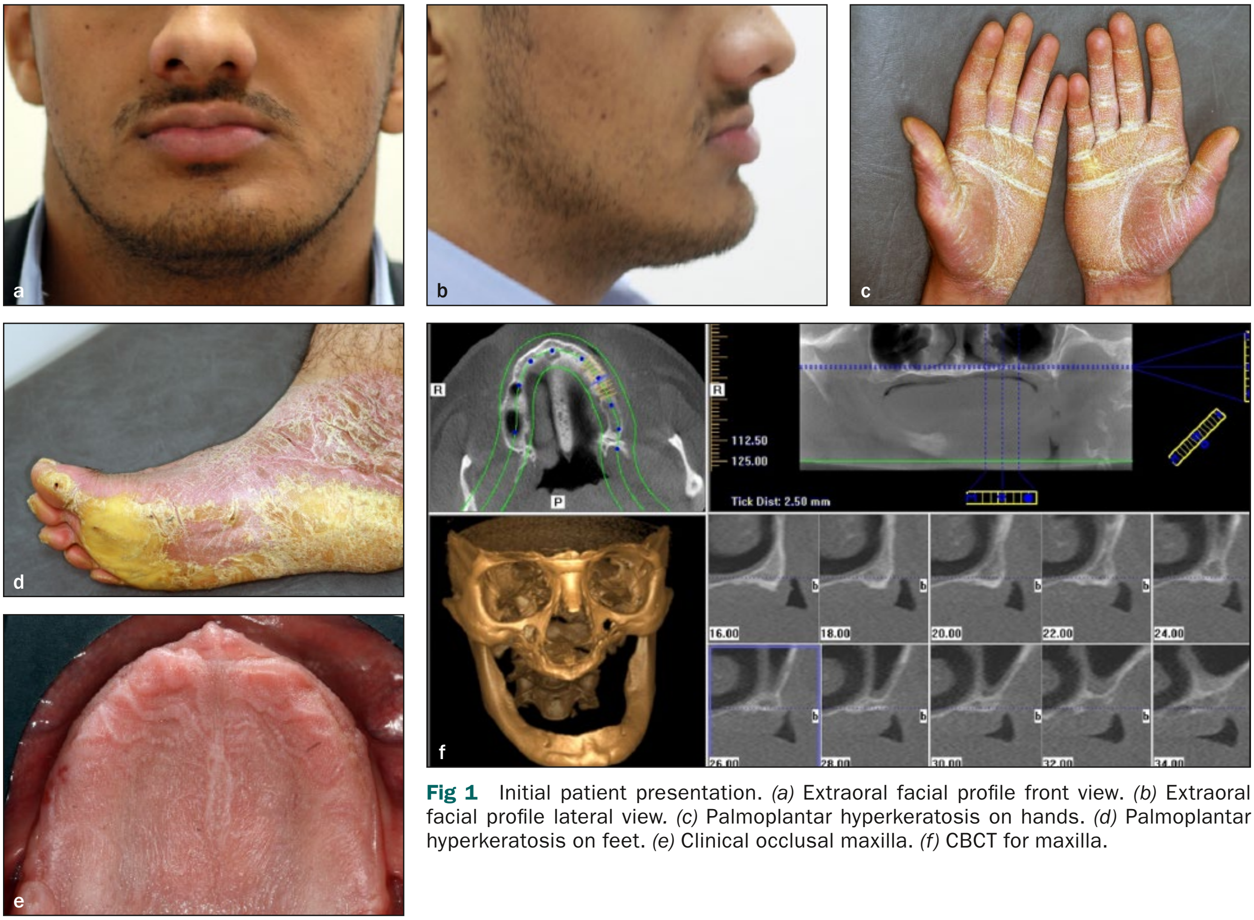
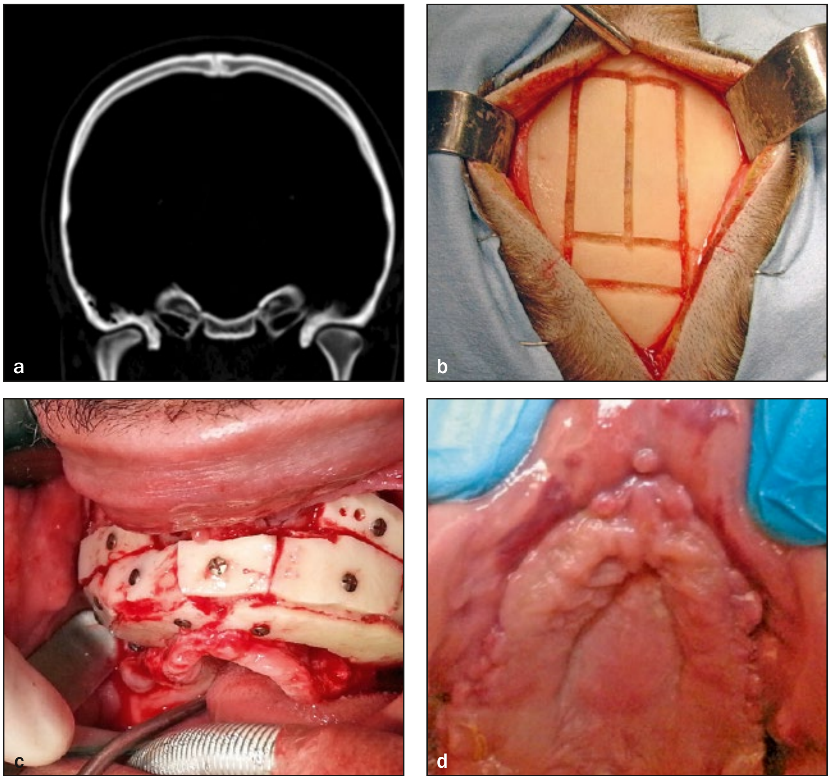
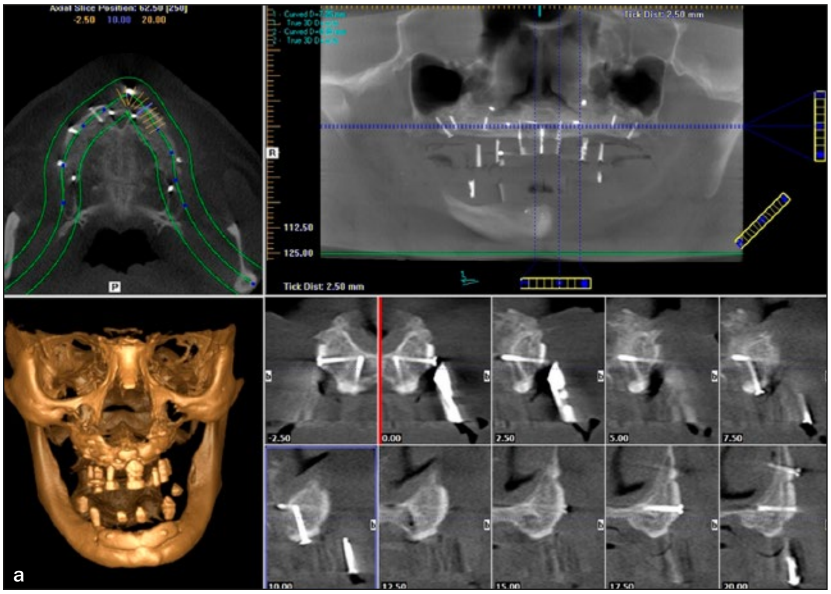


Fig 1 Initial patient presentation. *(a)* Extraoral facial profile front view. *(b)* Extraoral facial profile lateral view. *(c)* Palmoplantar hyperkeratosis on hands. *(d)* Palmoplantar hyperkeratosis on feet. *(e)* Clinical occlusal maxilla. *(f)* CBCT for maxilla.

Fig 2 *(a)* Medical CT scan of skull thickness. *(b)* Donor site preparation of bone strips. *(c)* Recipient site with graft strips secured using fixation screws. *(d)* Two weeks postsurgical maxilla.





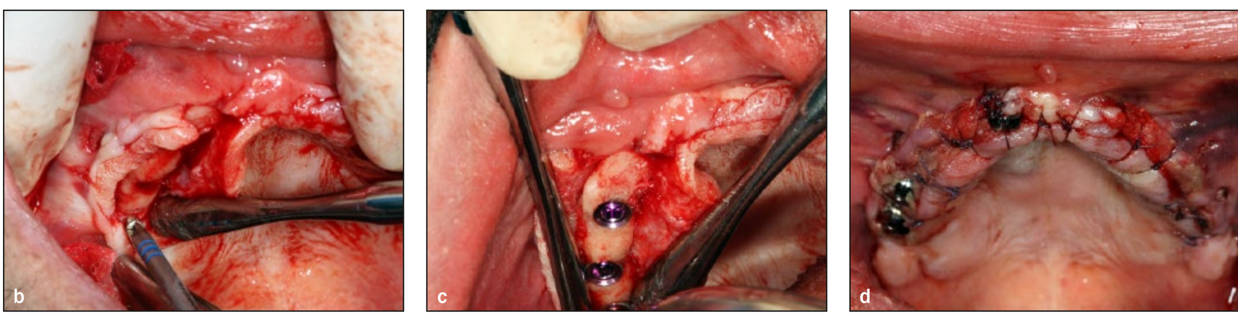


Fig 3 *(a)* CBCT 3 months after maxilla graft. *(b)* Fixation screw removal. *(c)* Implant placement. *(d)* Clinical view showing closure after implant placement in the maxilla.

Implant Surgery

An additional CBCT using a radiographic guide was taken, demonstrating adequate bone quantity and quality for dental implant placement (Fig 3a). The 3-month follow-up demonstrated excellent graft integration, and the patient was cleared for dental implant placement with B.M.K. On the day of implant surgery, the patient rinsed with a 0.12% chlorhexidine gluconate solution for 1 minute presurgically (Hi-Tech Pharmacal Co). Local anesthesia was administered using 2% lidocaine with 1:100,000 epinephrine (Empi). Using a surgical guide, the sequential implant osteotomy was initiated through the soft tissue without flap elevation to mark the implant sites. A midcrestal incision and full-thickness facial flap was reflected maintaining adequate keratinized gingiva around dental implants. The surgical fixation screws were removed, and osteotomy preparation continued according to the implant company surgical protocol (Figs 3b and 3c). Nine implants (Legacy 3, Implant Direct) were placed in the maxilla and six in the mandible, and flaps were approximated for closure using 4-0 resorbable sutures (Vicryl, Medline Industries) (Fig 3d).

RESULTS

The implants were allowed to heal for an additional 3 months, and healing abutments were placed (Fig 4a). Thereafter, implants were torqued with good osseointegration. Impressions were made, and an implant-supported prosthesis was fabricated and connected to the implants (Figs 4b and 4c). The 1-year follow-up showed well-osseointegrated implants with good function with esthetic smile, and the patient was satisfied with the treatment outcomes (Fig 4d). The patient continued to be seen every 4 to 6 months for periodontal and prosthesis maintenance. The results were satisfactory at the 12-month follow-up. This is the first reported case of cranium grafting for endosseous implant-supported full-arch prostheses in a PLS patient.

DISCUSSION

Patients affected by PLS have to manage severe oral disability from a very young age. This can negatively affect growing children socially, esthetically, and

Fig 4 *(a)* Panoramic radiograph after implant placement. *(b)* Intraoral facial view of implant-supported prostheses. *(c)* Lateral cephalogram of implant-supported prostheses. *(d)* Esthetic smile at 1-year follow-up.

physically. The present patient presented very frustrated with the failure of three previous removable prostheses and felt unable to function adequately in his day-to-day life. The reasons for severe rapidly progressing periodontitis in PLS patients have not been fully elucidated; however, the possible mechanisms have been classified into three domains: immunologic, microbiologic, and genetic.[19] From the immunologic perspective, neutrophils have displayed impaired chemotaxis, phagocytosis, and bactericidal activities.[20,21] Microbiologically, the presence of *A actinomycetemcomitans* has been noted in periodontal pockets of PLS patients, and could be considered as a triggering factor.[6,22] This finding has been expanded upon by Clerehugh et al, who identified a broader range of putative periodontopathogens with presence in the oral cavity of PLS-affected individuals, including *F nucleatum* and *P gingivalis*.[23] The investigation of genetic factors has led to the hypothesis that inactivation of the cathepsin-C gene is primarily responsible for abnormalities in the skin development and periodontal disease progression in PLS patients.[4] This gene is also mutated in two related conditions: Haim-Munk syndrome and aggressive periodontitis. A clinical manifestation common to all of these disorders is early onset, severe, and rapidly progressing periodontal destruction.

Implant dentistry has broadened the treatment options for severely atrophic arches unable to support, retain, or stabilize traditional removable prostheses. Autogenous bone harvested from the calvarium has been reported to provide adequate bone volume and contours to allow for predictable vertical and horizontal bone augmentation.[17] The use of cranium bone generally results in less morbidity compared with grafts taken from the tibia or iliac crest.[16] In a case series by Restoy-Lozano et al, a mean vertical bone gain of 5.04 ± 1.69 mm was obtained in 10 patients.[17] Provided that adequate bone volume is present or can be achieved, osseointegrated implants provide an alternative with major improvements in the long-term prognosis for oral rehabilitation.[15,24] A growing body of evidence continues to suggest that PLS patients should be considered candidates for full-arch implant-supported prostheses.

CONCLUSIONS

Although the long-term stability of PLS cases treated with implant-supported prostheses has not been established, the present case demonstrates that despite several failed attempts at prosthetic rehabilitation, a potential treatment option for the restoration of a severely atrophic arch consists of an interdisciplinary approach to treatment with autogenous calvarium grafts followed by endosseous implant placement and implant-supported prosthesis fabrication. Treatment planning may appear daunting for cases complicated by severe arch atrophy secondary to PLS, necessitating clear communication and partnership between members of the dental team to ensure proper and logical treatment progression. This case was managed using an interdisciplinary approach, resulting in a treatment outcome that met patient expectations, and led to a significant improvement to self-image and confidence in social settings.

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