

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/jpfa

# Papillon–Lefèvre syndrome – The prosthodontic management

*Rathika Rai, Arun Kumar*, Varun Deshpande*

*Department of Prosthodontics, Thai Moogambigai Dental College and Hospital, Chennai, India*

*Keywords:*
Palmoplantar hyperkeratosis
Floating in air appearance
Implant-supported prostheses
Locator attachments

ABSTRACT

Papillon–Lefèvre syndrome is a rare autosomal recessive genetic disorder. This is characterized by palmar-plantar hyperkeratosis, with rapidly progressive periodontitis and premature loss of both deciduous and permanent teeth. This article presents the prosthodontic rehabilitation of a patient with Papillon-Lefèvre syndrome with implant-supported over denture using two dental implants in mandible.

Copyright © 2014, Pierre Fauchard Academy (India Section). Publishing Services by Reed Elsevier India Pvt. Ltd. All rights reserved.

## 1. Introduction

Papillon–Lefèvre syndrome (PLS) is a very rare autosomal recessive disorder characterized by palmoplantar hyperkeratosis and severe early onset periodontitis, affecting the primary and permanent dentition. This syndrome was first reported and described by two French physicians, Papillon and Lefèvre in 1924 on two children with skin lesions on the hands and feet. These children experienced severe periodontal disease and the premature loss of the primary and secondary dentitions. The prevalence of PLS has been reported as 1–4 per million.[1] Radiographic evaluation will show teeth "floating in air appearance". The etiopathogenesis of the syndrome is relatively unknown. It can be of immunologic, genetic, or possible bacterial etiologies.[2] Genetic analysis suggests that the disorder may result from mutations of a gene that regulates production of an enzyme known as cathepsin C, which are seen more in osteoclasts.[3] The gene is located on the long arm (q) of chromosome 11. Cathepsin C participates in both intracellular and extracellular cleavage of proteins and activation of serine proteases in immune and inflammatory cells.

It is characterized by palmoplantar keratoderma (thickening of the stratum corneum of the skin on the palms of the hands and soles of the feet) and periodontitis followed by the premature shedding of both primary and permanent teeth. Palmoplantar keratosis typically begins from 1 to 4 years of age.[4] It can also manifest with excessive sweating (hyperhidrosis), the growth of fine body hair and the development of dirty colored skin on the affected parts, calcifications of the falx cerebri of the dura mater, as well as other areas of the brain have been reported. From a dental standpoint, young patients with PLS have juvenile periodontitis, severe destruction of the alveolar bone as early as two years of age in both primary and permanent dentitions.

Conventional periodontal therapy generally fails in patients with PLS, and rapid progression of periodontitis frequently results in a severe alveolar bone loss. Microbiological studies of the oral microflora of patients with PLS have shown that the predominant organisms in the periodontal sites are Gram-negative anaerobic rods. Reduced neutrophil phagocytosis, bacterial infection, and impaired reactivity to T- and B-cell mitogens could be the reason for the periodontal disease and increased susceptibility to infection.[5] Conventional periodontal treatment usually fails in PLS patients,

* *Corresponding author*. 21, SMV Koil Street, Triplicane, Chennai 600005, India. Tel.: +91 9940578990.
E-mail addresses: dr_arun_bds@yahoo.co.in, dr.arun.bds25@gmail.com (A. Kumar).
http://dx.doi.org/10.1016/j.jpfa.2014.03.002
0970-2199/Copyright © 2014, Pierre Fauchard Academy (India Section). Publishing Services by Reed Elsevier India Pvt. Ltd. All rights reserved.

resulting in the rapid progression of periodontitis and destruction of alveolar bone. Early extraction of periodontally involved permanent teeth has been considered to preserve supporting alveolar bone. In addition, the periodontal disease may be arrested by improving oral hygiene, extracting severely diseased teeth, scaling, administering systemic antibiotics, and providing long-term antimicrobial irrigation.

Previously the treatment approach was extraction and conventional prosthodontic rehabilitation. Recently, endosseous dental implant-supported prostheses have been considered as an alternative method. Implant-supported prostheses enhance the support, stability, and retention of prostheses. Ullbro et al,[6] Woo et al,[7] and Etöz et al[8] have shown that the implant therapy is an option for successfully restoring edentulism in patients with Papillon–Lefèvre syndrome. Excessive loss of alveolar bone support may jeopardize the placement of implants in PLS cases. In such situations, either bone augmentation or use of short implants may be considered.

This article presents the rehabilitation of a patient with Papillon–Lefèvre syndrome with dental implants.

## 2. Case report

A 26-year-old male patient reported to the department of prosthodontics, Thai Moogambigai Dental College and Hospital, Chennai with the chief complaint of unaesthetic appearance and difficulty in eating because of constant exfoliation of teeth (Fig. 1). The patient's medical history revealed that before 6 years he has been diagnosed with Papillion–Lefèvre syndrome. He was treated with oral Isotretinoin, three times daily to improve the dermatological problem.

Patient gave a dental history of premature exfoliation of his teeth from childhood. Family history revealed that parents had a consanguineous marriage. His sibling was not affected by the disease. On general examination, the patient was thin built with dry scaly keratotic plaques over the skin of his hands and legs (Fig. 2). On intraoral examination, revealed partially edentulous condition with remaining teeth were of grade III mobile and grade II mobile horizontally impacted 38 (Fig. 3). Radiographic examination revealed the classic presentation of teeth "floating in air appearance" (Fig. 4).



**Fig. 1 – Pretreatment extraoral photograph.**

Considering Muller De Van statement[9] "The preservation of that which remains is of utmost importance and not the meticulous replacement of that which has been lost", extensive surgical options like bone augmentation was avoided. Considering his age and the prognosis of the teeth, a decision was made to extract all the teeth (Fig. 5). To restore function and esthetics, implant-supported over denture for his mandible was considered as the treatment option.

A preoperative Panoramic radiograph was taken before implant placement. Two implants of 3.3 × 10 mm and 3 × 11.5 mm (Fig. 6) were placed in the B and D position of the mandible for an OD1 overdenture treatment option. The implant placement surgery was done in the operating room under local anesthesia (Fig. 7). An immediate postoperative Panoramic radiograph was taken. The patient was placed on antibiotic regimen and 2% chlorhexidine rinse for 2 weeks. He was followed-up weekly for the first month and then at regular intervals. The postsurgical recovery period was uneventful with no chief complaint or complications. The lower complete denture was first relined periodically with soft liner (GC Corporation, Tokyo, Japan) to avoid immediate loading of the implant during osseointegration. After 6 months clinical and radiographical examination done to determine osseointegration (Fig. 8). The criteria for success were the absence of mobility, the absence of radiographic gap in the bone-implant interface, and the absence of pain or infection at the peri-implant area. The implants were subsequently uncovered and healing abutments were placed. The patient was under 2% chlorhexidine rinse for 2 weeks.

The first phase of prosthodontic treatment, Primary impressions were made with irreversible hydrocolloids (Prime-chrome, Prime Dental Products Pvt. Ltd.) for maxillary and mandibular arch after 2 weeks. An open tray implant level impressions were made with addition silicone (Aquasil, Dentsply). Facebow records were made and routine clinical procedures were followed to obtain jaw relation records and the casts were mounted on the semi adjustable hanau articulator. Teeth arrangement was done with characterization to give a refreshing smile. During try-in the esthetic aspects were discussed with the patient and the patient's approval was obtained. The lower overdenture was modified with the O–ring-type of locator attachments (Figs. 9 and 10) and stabilized through these implants to attain PM 6 movement. Dentures were processed in high impact heat polymerized acrylic resin (Trevalon, Dentsply). After finishing and polishing the dentures were inserted (Figs. 11 and 12). Post-insertion instructions were given to the patient. Necessary adjustments were done in recall visits. The patient was under periodical visits.

## 3. Discussion

Papillon-Lefèvre syndrome is characterized by palmar-plantar hyperkeratosis and periodontitis occurring in childhood after



Fig. 2 – Dry scaly keratotic plaques over the skin of hands and legs.



Fig. 3 – Pre-extraction intraoral photograph.



Fig. 5 – Post-extraction intraoral photograph.



Fig. 4 – Pre-extraction panoramic radiograph.

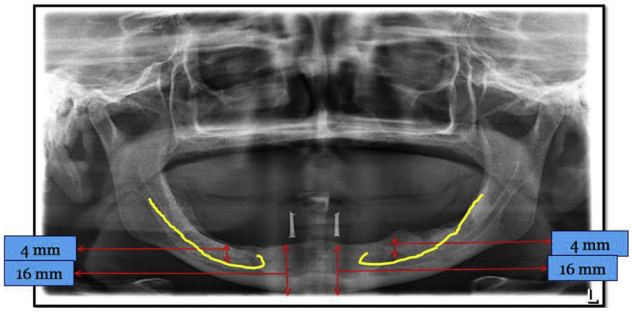



Fig. 6 – Radiographic interpretation of available bone.

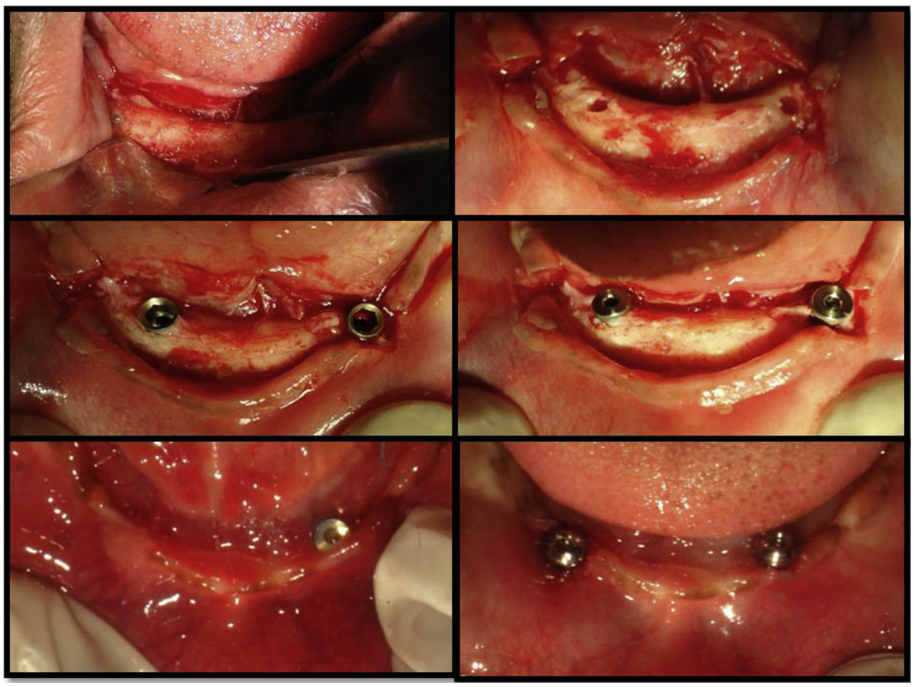


**Fig. 7 – Osteotomy site.**

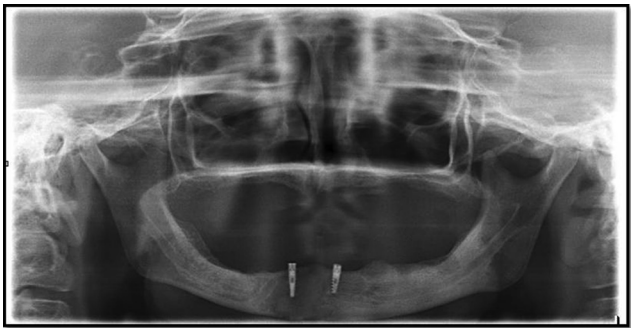


**Fig. 8 – Radiographic evaluation of osseointegration.**



**Fig. 10 – Denture with O ring type attachment.**

eruption of the deciduous teeth. The population prevalence of Papillon–Lefèvre syndrome is reported to be 1 case in every 4 million people. This patient had parent consanguinity and this is similar to a review reported by Gorlin.[10]

The pathogenesis of this syndrome is still not understandable. Etiology can be of Immunologic, genetic, and microbiologic factors. As a genetic factor cathepsin C gene is associated with Papillon-Lefèvre syndrome. The cathepsin C gene is seen in the epithelial regions commonly affected by Papillon–Lefèvre syndrome, such as the palms, soles, knees,



**Fig. 9 – Ball attachments.**



**Fig. 11 – Post-operative intraoral photograph.**



**Fig. 12 – Post-operative extraoral photograph.**

and keratinized oral gingiva.[11] In addition, it is expressed in various immune cells including polymorphonuclear leukocytes and macrophages, as well as their precursors.[12]

The fabrication of upper and lower complete dentures must restore form and function for this patient. However, because of the inadequate and continual loss of bone support in the mandible, the stability of the lower denture was compromised. Hence it was concluded that the viable treatment option was implant-retained overdenture following Ullbro et al[6] and Woo et al,[7] reports that the successful outcome of implant treatment in patients with Papillon–Lefèvre syndrome is achievable. Etöz et al[8] showed that patients with Papillon-Lefèvre syndrome can be safely managed with dental implants in atrophic mandibles, instead of invasive pre-prosthetic applications such as bone augmentation, nerve lateralization, or alveolar distraction, to avoid possible complications. Abdullah et al[13] has shown a one-year follow-up of the case showing functionally and esthetically stable dental implant with no signs of infection or bone loss.

## 4. Conclusion

Papillon–Lefèvre syndrome patients can be effectively treated using implants as the chief complaint of these patients was dentures with compromised retention. This case report has shown successful 1-year follow-up of implant osseointegration and alveolar bone preservation in a Papillon–Lefèvre syndrome patient wearing a functional implant-retained overdenture. This treatment option provides a viable option for restoring edentulism in young individuals suffering from this disorder and improving the quality of life.

## Conflicts of interest

All authors have none to declare.

## REFERENCES


1. French D, Scott H, Overall CM. Papillon–Lefevre syndrome associated early onset periodontitis: a review and case study. *J Can Dent Assoc*. 1995;61:432–438.
2. Dalgic B, Bukulmez A, Sari S. Eponym: Papillon–Lefevre syndrome. *Eur J Pediatr*. 2011;170:689–691.
3. Hakeda Y, Kumegawa M. Osteoclasts in bone metabolism. *Kaibogaku Zasshi. J Anat*. 1991;66:215–225.
4. Bach JN, Levan NE. Papillon-Lefèvre syndrome. *Arch Dermatol*. 1968 Feb;97:154–158.
5. Robertson KL, Drucker DB, James J, Blinkhorn AS, Hamlet S, Bird PS. A microbiological study of Papillon–Lefevre syndrome in two patients. *J Clin Pathol*. 2001;54:371–376.
6. Ullbro C, Crossner CG, Lundgren T, Stalblad PA, Renvert S. Osseointegrated implants in a patient with Papillon–Lefevre syndrome. A 4 1/2-year follow up. *J Clin Periodontol*. 2000;27:951–954.
7. Woo I, Brunner DP, Yamashita DD, Le BT. Dental implants in a young patient with Papillon–Lefevre syndrome: a case report. *Implant Dent*. 2003;12:140–144.
8. Etöz OA, Ulu M, Kesim B. Treatment of patient with Papillon–Lefevre syndrome with short dental implants: a case report. *Implant Dent*. 2010;19:394.
9. DeVan M. The nature of the partial denture foundation: suggestions for its preservation. *J Prosthet Dent*. 1952;2:210–218.
10. Gorlin RJ. Of palms, soles, and gums. *J Med Genet*. 2000;37:82.
11. Cagli NA, Hakki SS, Dursun R, et al. Clinical, genetic, and biochemical findings in two siblings with Papillon–Lefèvre syndrome. *J Periodontol*. 2005;76:2322.
12. Sollecito TP, Sullivan KE, Pinto A, et al. Systemic conditions associated with periodontitis in childhood and adolescence (A review of diagnostic possibilities). *Med Oral Patol Oral Cir Bucal*. 2005;10:142.
13. Abdullah. Oral rehabilitation of a case of Papillon–Lefevre syndrome with dental implants. *Saudi Med J*. 2013;34:424–427.