BULLETIN OF STOMATOLOGY AND MAXILLOFACIAL SURGERY
Volume 20, Issue 4

DOI:10.58240/1829006X-2024.4-66

CLINICAL ARTICLE

Full-mouth dental implant rehabilitation for a patient suffering from Papillon-Lefevre syndrome

Elie Maalouf[1], Wadih Nassif[2], Eliano Nakhoul[1], Fady Abillamaa[1], Georges Aoun[3*]

[1]Department of Periodontology, Faculty of Dental Medicine, Lebanese University
[2]Department of Prosthodontics, Faculty of Dental Medicine, Lebanese University
[3]Department of Oral Medicine and Maxillofacial Radiology, Faculty of Dental Medicine, Lebanese University

[*]**Corresponding author**: Professor Georges Aoun, Department of Oral Medicine and Maxillofacial Radiology, Faculty of Dental Medicine, Lebanese University, Beirut, Lebanon. E-mail: dr.georgesaoun@gmail.com, ORCID ID: http://orcid.org/0000- 0001-5073-6882.

Received: *Aug 1st, 2024;* ***Accepted:*** *Aug 30th, 2024;* ***Published:*** *Sep 15th, 2024.*

Abstract

Papillon-Lefevre syndrome (PLS) is an autosomal recessive disorder that affects the primary and permanent dentitions. It is characterized by severe periodontitis. Titanium implants have emerged as a significant therapeutic option in recent years for both tooth replacement and oral rehabilitation in individuals with PLS. Due to the severe bone loss, these patients may require bone grafting procedures, sinus lifts, and the alveolar ridge splitting technique (ARST) in order to properly position the implants. This report aims to demonstrate how a patient with PLS can be successfully managed for full-mouth rehabilitation with dental implants and hybrid prosthesis.
Periodontists and prosthodontists should combine their efforts for an optimal customized treatment plan.
Dental implants have been successfully used to restore functionally and esthetically the patient's mouth.

Keywords: Papillon-Lefevre syndrome; dental implants; bone grafting; sinus lift; alveolar ridge splitting technique; full-mouth rehabilitation.

Introduction

In 1924, Papillon and Lefevre published the first description of the Papillon-Lefevre syndrome (PLS). It is an autosomal recessive disorder caused by a point mutation in the cathepsin C gene that affects the primary and permanent dentitions. It is characterized by generalized palmoplantar hyperkeratosis and a quickly developing and severe periodontitis.
PLS has a reported prevalence of 1-4 cases per million. Usually, the eruption of the main teeth coincides with the onset of the disorder. No race is significantly affected; boys and girls are equally afflicted. After the eruption of the primary teeth, the gingiva becomes irritated. Most affected kids lose their primary teeth prematurely, and the periodontium typically deteriorates quickly after this. Gingiva appears to return to its typical state following the main dentition's exfoliation. After the permanent teeth erupt, the aggressive, inflammatory periodontal process recurs, and most or all of the permanent dentition is lost throughout adolescence.[1]
Because of the movement of the teeth, chewing is extremely uncomfortable. Usually, halitosis is noticeable. Additionally, there has been evidence of regional lymphadenopathy tenderness.
In advanced cases, radiographic imaging shows significant alveolar bone loss, and teeth seem to be "floating-in-air." While unerupted teeth often grow properly in their bone crypts, occasionally they adopt an unusual posture and have partially developed roots.[2,3]
Conventional periodontal therapy is usually not effective in treating patients with PLS.[4] In order to preserve the alveolar bone, early extraction of

permanent teeth has been suggested.[5]

Nowadays, dental implants constitute a key therapeutic option for tooth replacement and oral rehabilitation in patients with large edentulous.[1]

This article reports the successful management of a patient with PLS for full-mouth rehabilitation with dental implants and hybrid prosthesis.

Case Presentation

A 21-year-old non-smoking female patient affected by PLS since childhood was referred by her general dentist to our Oral and Maxillofacial Surgery specialized clinic to restore functionally and esthetically her mouth. Extraoral clinical exam revealed diffuse palmoplanter hyperkeratosis (Figure 1).

Figure1. Extraoral clinical photos showing hyperkeratotic plaques on the hands and feet of the patient

An old panoramic radiograph showed severe bone loss and many radiolucent periapical images (Figure 2).

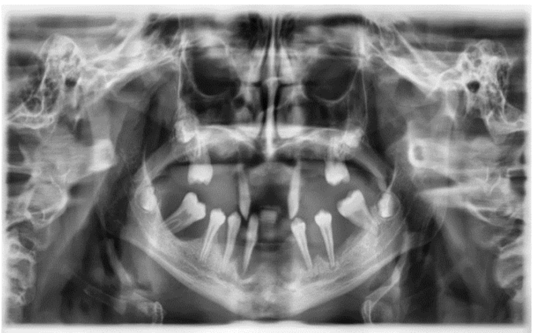

Figure 2. Old panoramic x-ray extraction showing the absence of several teeth and the presence of severe bone loss on all remaining teeth with radiolucent periapical images

The actual intraoral clinical exam showed a complete edentulous mouth except for tooth #45. A good amount of keratinized gingiva was present on buccal maxillary sites, while it was deficient on buccal mandibular sites (Figure 3).

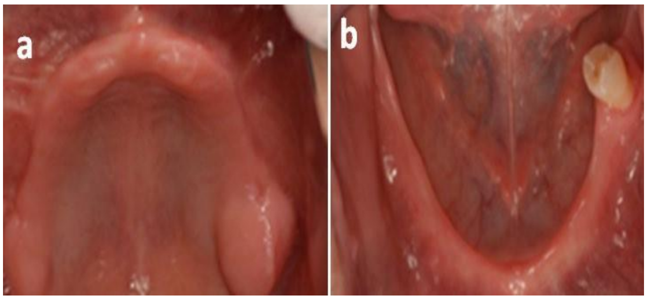

Figure 3. (a) Occlusal intraoral photo of the upperarch; (b) occlusal intraoral photo of the lower arch

The cone beam computed tomography (CBCT) para-axial cuts showed insufficient bone height and width everywhere except for the lower anterior region (Figure 4).

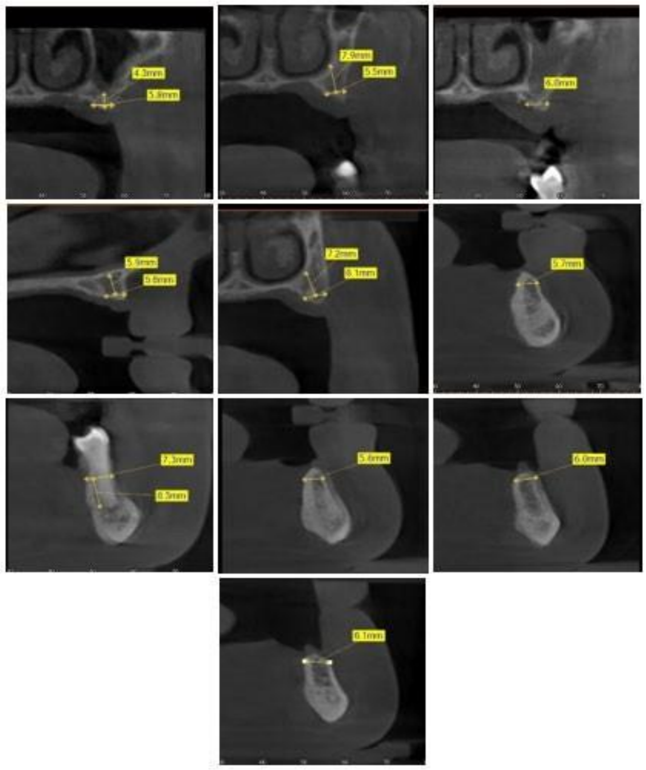

Figure 4. Para-axial cuts on lower and upper arches; note the insufficient bone height and width on all sites except for the lower anterior region.

Elie Maalouf, Wadih Nassif, Eliano Nakhoul, Fady Abillamaa, Georges Aoun. Full-mouth dental implant rehabilitation for a patient suffering from Papillon-Lefevre syndrome Bulletin of Stomatology and Maxillofacial Surgery. 2024;20(4): doi: 10.58240/1829006X-2024.4-66

In light of these findings, the treatment plan was to extract tooth #45, place five implants on the mandible, and place another five implants on the maxilla with the ridge splitting technique and an internal sinus lift on implant sites #13 and #15.
After several months of surgery, the final restoration will be placed. The treatment plan was approved by the patient.
Under local anesthesia using 4% articaïne HCl (1:100,000 epinephrine), a midcrestal incision was made on the upper arch using a 15c blade. A complete exposure of the alveolar bone was provided by a full-thickness mucoperiosteal flap elevation.
On the right side, bone width was insufficient for implant placement, and residual bone height (RBH) was only 4 mm, as shown on the CBCT.
Responding to this situation, a split thickness technique and internal sinus floor elevation were made simultaneously with implant placement on #15 and #13.
Using a piezo instrument, one horizontal and two vertical bone splits were made. Using the round bur, the first implant site #13 was marked, and then twist drills (ø 2.2, ø 2.8, and ø 3.5 mm) were used consecutively at 11 mm height.
On site #15 (at 12 mm from site #13), where the bone height is 4 mm, an implant bed was prepared 1 mm below the sinus floor using ø 2.2, ø 2.8, and ø 3.5 mm tapered twist drills consecutively respecting the parallelism using the gauge placed in the implant site.
Using a ø 3.5 mm osteotome, the sinus floor was elevated according to the direct osteotome sinus floor elevation technique. The Valsalva test was done in order to control the continuity of the membrane.
Five to six scoops (equivalent to 4 to 5 mm of membrane elevation) of the grafting material (xenograft bovine bone substitute BioOss Githlesh) were progressively condensed and pushed by the osteotome, which remains within the 4 mm of crestal height; the elasticity and membrane integrity were controlled repetitively.
A 4.1*10 mm Roxolid Straumann® tissue level implant (Straumann AG, Basel, Switzerland) was placed with the treated surface at 1 mm below the alveolar crest on implant site #13. Another 4.1*8 mm Roxolid Straumann® tissue level implant (Straumann AG, Basel, Switzerland) was placed with the treated surface at 1 mm below the marginal alveolar crest on implant site #15, and appropriate cover screws were placed.
Gaps between implants and bone were filled with a xenograft bone substitute, BioOss Githlesh (Figure 5).

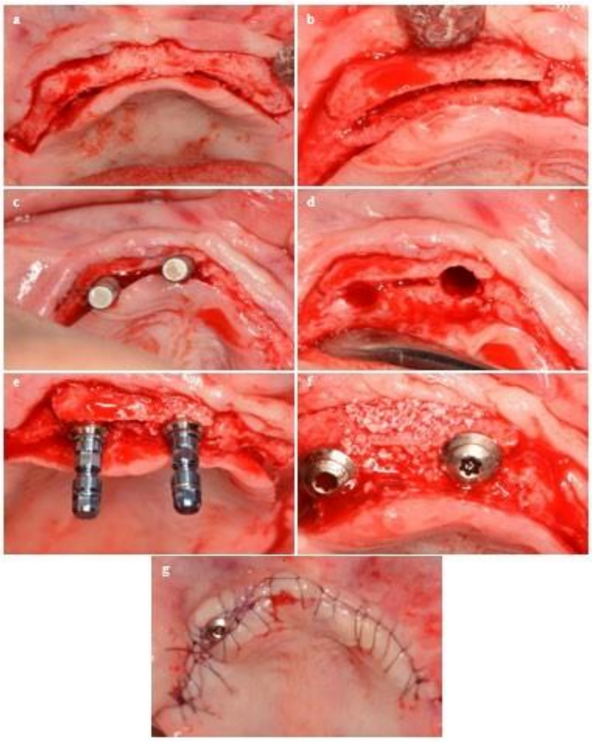

Figure 5. (a) Occlusal intraoral photo of the upper arch during full thickness flap elevation; (b) one horizontal and two vertical bone splits during alveolar ridge splitting technique; (c) two gauges placed in the implant sites to check the parallelism before implants placement; (d) occlusal view of the surgical sites showing the two holes after implant sites drilling and the bone splits used to increase bone width; (e) the two implants placed on the surgical sites #13 and #15, note the parallelism between these two implants; (f) occlusal view of the two implants placed, note the xenograft substitutes used to fill the gap between bone plates after splitting; (g) occlusal view of the upper arch showing the continuous sutures.

The patient was unable to continue the surgery, so the flap was readapted and sutured using continuous and interrupted sutures with PGA 4/0 and monofilament 5/0.
After three months, the surgery was continued on the upper arch, and three more implants were placed on implant sites #21, #23, and #25, a 4.1*10 mm Roxolid Straumann® tissue level implant (Straumann AG, Basel, Switzerland) (Figure 6).

On the lower arch, tooth #45 was extracted, and five implants were placed on implant sites #45 (immediate placement), #42, #41, #31, and #33, a 4.1*10 mm Roxolid Straumann® tissue level implant

(Straumann AG, Basel, Switzerland) (Figure 7).

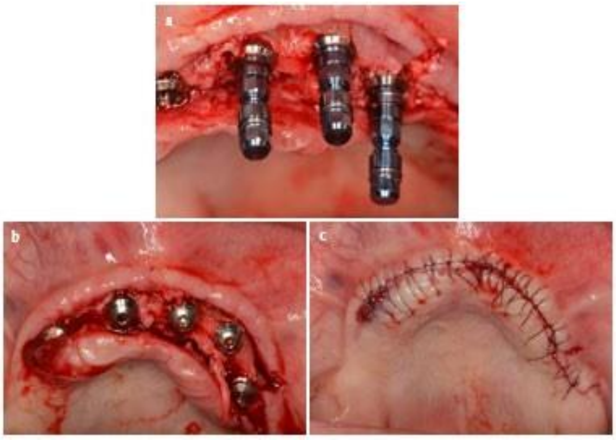

Figure 6. (a) Close-up photo showing the parallelism between the implants placed on surgical sites #21, #23, and #25; (b) occlusal view of the upper arch before flap closure; note the five tissue-level implants placed in order to support the prosthesis; (c) continuous sutures used to close the flap tightly.

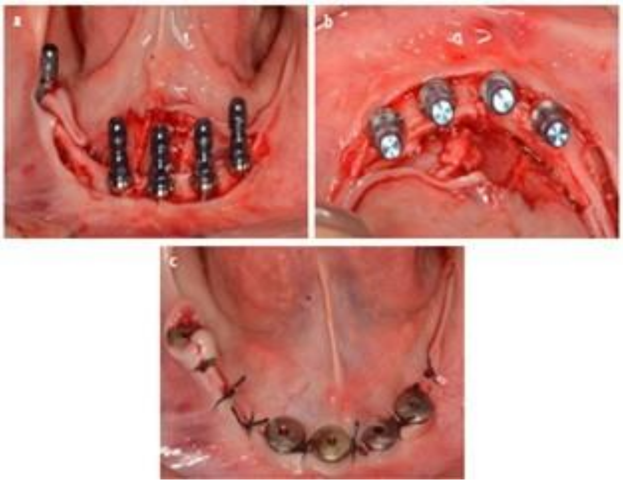

Figure 7. (a) Intraoral photo showing the parallelism between the implants placed on the lower surgical sites #45, #42, #41, #31, and #33; (b) occlusal view of the lower implants; note the ideal bucco-lingual position of these implants; (c) sutures placed between the implants to close the flap.

The patient was told to apply ice packs every five minutes for the first three hours following each procedure. It was advised to take 2 g of amoxicillin and clavulanic acid daily for 7 days in order to prevent infection at the surgery site. In cases of pain, two 500 mg paracetamol tablets were advised every six hours.

The 0.12% CHX mouthwash was administered for 14 days after surgery, starting on the second post-operative day. During the planned follow-ups, there were no postoperative adverse effects seen, and the healing process proceeded without any problems. The patient was seen eight months after the last surgery. The clinical exam revealed a healthy gingival aspect on the implant sites; the buccal and occlusal views show a normal-shaped, orange peel-appearance gingiva (Figure 8).

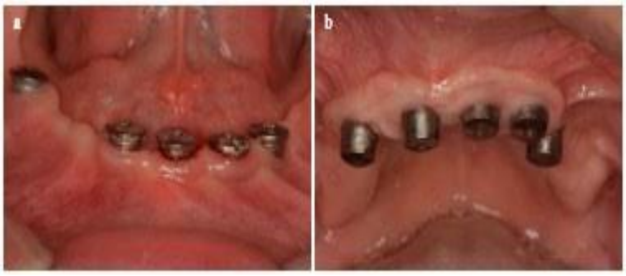

Figure 8. (a) Close-up photo of the five lower implants with their healing abutments; note the good amount of keratinized, pink, and healthy gingiva around the implants; (b) close-up photo of the five upper implants with their healing abutments; note the healthy soft tissue in this region.

The final impression was taken using splinted open tray copings impression. Vertical dimension and teeth mounting were tried on to ensure esthetic and patient comfort (Figure 9).

Figure 9. (a) Close-up photo of the five upper coping impressions during the final impression; note the red

acrylic used to splint all the components; (b) intraoral photo showing the five lower coping impressions splinted together; (c) photo of the lower gypsum model showing the five implant analogs; (d) photo of the upper gypsum model showing the five implant analogs; (e) photo of the two models articulated with teeth setting up; (f) try-in of the two prosthesis in the patient's mouth to verify esthetics, occlusal, and vertical dimensions.

Finally, two hybrid prostheses were placed on the upper and lower arches using multiunits abutments (Figure 10).

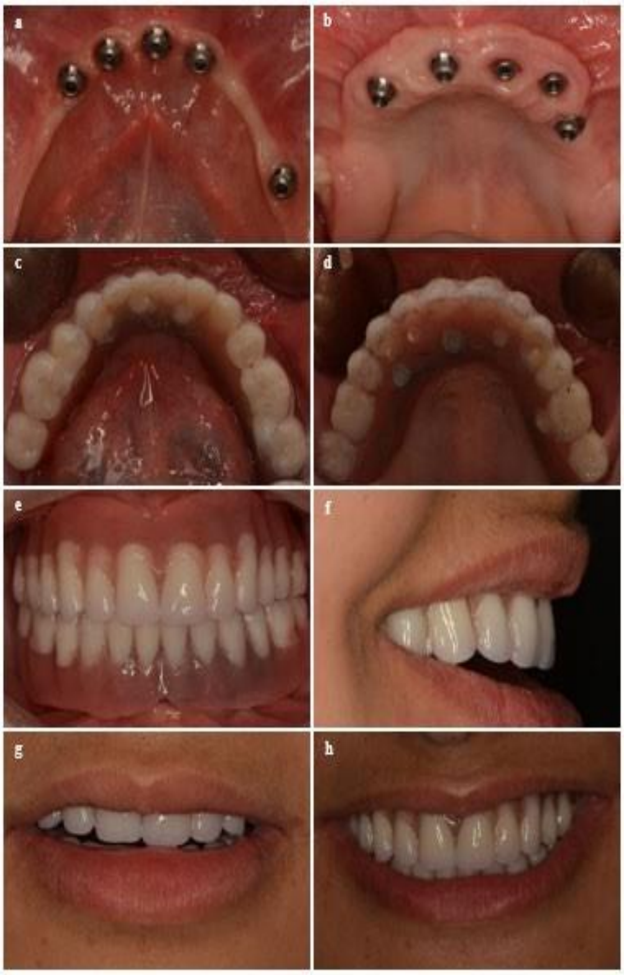

Figure 10. (a) Intraoral occlusal view photo of the lower arch showing the five multiunits screwed on the implants; (b) intraoral occlusal view photo of the upper arch showing the five multiunits screwed on the implants; (c) intraoral occlusal view photo of the lower hybrid prosthesis after insertion; (d) intraoral occlusal view photo of the upper hybrid prosthesis after insertion; (e) frontal intraoral view photo of the upper and lower prosthesis in occlusion position; note the harmony of the marginal artificial gingiva and between teeth sizes; (f) lateral extraoral view of the smile; note the ideal position of the artificial gingiva with the upper lip; (g) extraoral view photo in rest position showing a part of the anterior teeth; (h) extraoral view photo of the full smile of the patient showing a perfect pink and white esthetics.

Two weeks after prostheses insertion (one year after last surgery), a panoramic radiograph was done. This latter showed the absence of any abnormal radiolucency or any bone loss on all implant sites, and it also revealed the perfect fit of all the abutments on their implant platform (Figure 11).

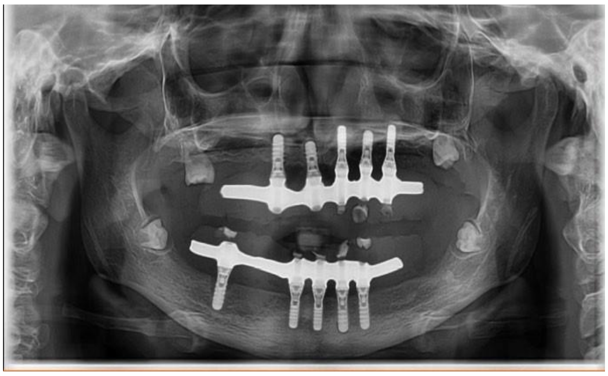

Figure 11. Panoramic x-ray done one year after last surgery; note the absence of any bone loss around upper and lower implants, absence of any abnormal radiolucency, and the perfect fitting of the two hybrid prostheses.

Discussion

One significant sign of PLS is periodontitis, and in most cases, conventional periodontal therapy is ineffective in stopping the chronic loss of attachment in these individuals. Antimicrobial medications such as erythromycin, tetracycline, penicillin, and amoxicillin-metronidazole have been proposed in addition to periodontal therapy. Nonetheless, there have been reports of controversial outcomes. Some patients experience tooth loss and continuous attachment loss. These patients require mouth repair because they suffer from partial or total edentulism in their teens.[6] For PLS patients, the conventional prosthetic treatment consists of an overdenture or complete denture; however, these devices have aesthetic and functional drawbacks that prompt patients to look for more comprehensive care. The primary complaints expressed by patients concerned the instability and retention of their mandibular prosthesis.

PLS sufferers can benefit from dental implants because they can give dental prosthesis the support, stability, and retention they need. Implants not only improve prosthesis stability and retention, but they help preserve the supporting bone and stop additional bone loss.[7]

In 2020, Atarbashi-Moghadam et al.[7] published a systematic review assessing oral rehabilitation of PLS patients by dental implants. It included 11 publications covering 15 cases treated by 136 implant placements.

Based on this review, three patients (20 implants) experienced implant failure. Two patients presented peri-implantitis (caused by inadequate oral hygiene and noncompliance with the maintenance program), and the third one presented a lack of osseointegration. In the three cases, the implant replacement was successful.

In our case, there was no implant failure after 2 years of the 10 implants placed in the maxilla and mandible. The multidisciplinary work (periodontist, prosthodontist, and dental technician), the good treatment plan, the well-experienced practitioners, and the good compliance of the patient with the oral hygiene instructions were definitely the reasons for this success.

To improve clinical outcomes in individuals with insufficient residual bone height, new approaches to maxillary sinus floor elevations are crucial. Moreover, to maintain appropriate implant placement and guarantee its survival, any horizontal alveolar bone deficiency must be corrected.[8]

In our case report, we performed the alveolar splitting technique and internal sinus lift to regenerate the bone horizontally and vertically in 2 implant sites in order to place the standard length and diameter implant (4.1*10) in an ideal position.

In another systematic review published by Bassetti et al.[9] in 2016 comparing the ARST and the expansion technique, it was reported that implants inserted using ARST have success rates that are similar to implants inserted in pristine bone without ARST. Nonetheless, compared to implants inserted in pristine bone without ARST, a somewhat more noticeable marginal bone loss might be anticipated, according to the few available data.[9]

In our case, ARST was used on the two implant sites (#13 and #15) in order to place 2 implants with a 4.1 mm diameter. The two implants did not show any complications or bone loss during the 2-year follow-up.

Conclusion

PLS is a highly uncommon inherited disorder that is autosomal and recessive in nature. This syndrome affects negatively patient's social, functional, aesthetic, and psychological characteristics.The affected patient must be managed with a multidisciplinary dental approach. Periodontist and prosthodontist should combine their efforts for an optimal customized treatment plan. Despite the periodontal damage resulted by this syndrome, dental implants became the most famous treatment to restore functionally and esthetically the patient's mouth.

Declarations

Ethics approval and consent to participate
Not applicable

Consent for publication
Written informed consent was obtained from the patient for publication of this case report.

Availability of data and materials
The data will be available on reasonable request from the corresponding author.

Competing interests
The authors declare no conflict of interest.

Funding
This research received no external funding

REFERENCES

1. Abdul NS, Dagriri LK, Shenoy M. Papillon-Lefévre Syndrome: A Rare Case Report and a Brief Review of Literature. Cureus. 2022;14(6):e26163.doi: 10.7759/cureus.26163.
2. AIBarrak ZM, Alqarni AS, Chalisserry EP, Anil S. Papillon-Lefèvre syndrome: a series of five cases among siblings. J Med Case Rep. 2016;10(1):260. doi:10.1186/s13256-016-1051-z.
3. Kleinfelder JW, Topoll HH, Preus HR, Müller RF, Lange DE, Böcker W. Microbiological and immunohistological findings in a patient with Papillon-Lefèvre syndrome. J Clin Periodontol. 1996;23(11):1032-1038.doi: 10.1111/j.1600-051x.1996.tb00533.x.
4. Leuenberger M, Fischer L, Mylonas A, Huber M, Di Domizio J, Gilliet M, et al. Papillon-Lefevre syndrome treated by acitretin: case report and cytokine profile. J Eur Acad Dermatol Venereol.

Elie Maalouf, Wadih Nassif, Eliano Nakhoul, Fady Abillamaa, Georges Aoun. Full-mouth dental implant rehabilitation for a patient suffering from Papillon-Lefevre syndrome Bulletin of Stomatology and Maxillofacial Surgery. 2024;20(4): doi: 10.58240/1829006X-2024.4-66

2022;36(3):e235-e238.doi: 10.1111/jdv.17772.

5. Ullbro C, Crossner CG, Nederfors T, Alfadley A, Thestrup-Pedersen K. Dermatologic and oral findings in a cohort of 47 patients with Papillon-Lefèvre syndrome. J Am Acad Dermatol. 2003;48(3):345-351. doi: 10.1067/mjd.2003.197.
6. Ullbro C, Crossner CG, Lundgren T, Stålblad PA, Renvert S. Osseointegrated implants in a patient with Papillon-Lefèvre syndrome. A 4 1/2-year follow-up. J Clin Periodontol.2000;27(12):951-954.doi: 10.1034/j.1600-051x.2000.027012951.x.
7. Atarbashi-Moghadam F, Atarbashi-Moghadam S, Kazemifard S, Sijanivandi S, Namdari M. Oral rehabilitation of Papillon-Lefèvre syndrome patients by dental implants: a systematic review. J Korean Assoc Oral Maxillofac Surg. 2020;46(4):220-227.doi: 10.5125/jkaoms.2020.46.4.220.
8. Mudalal M, Sun XL, Li X, Fang J, Qi ML, Wang J, et al. Minimally invasive endoscopic maxillary sinus lifting and immediate implant placement: A case report. World J Clin Cases. 2019;7(10):1234-1241. doi: 10.12998/wjcc.v7.i10.1234.
9. Bassetti MA, Bassetti RG, Bosshardt DD. The alveolar ridge splitting/expansion technique: a systematic review. Clin Oral ImplantsRes.2016;27(3):310-324.doi: 10.1111/clr.12537.

Elie Maalouf, Wadih Nassif, Eliano Nakhoul, Fady Abillamaa, Georges Aoun. Full-mouth dental implant rehabilitation for a patient suffering from Papillon-Lefevre syndrome Bulletin of Stomatology and Maxillofacial Surgery. 2024;20(4): doi: 10.58240/1829006X-2024.4-66