

# Oral rehabilitation of a case of Papillon-Lefevre syndrome with dental implants

Abdullah Al Farraj AlDosari BDS, DMSc.

## ABSTRACT

متلازمة بابيلون لي فيفر (بي أل أس) هو اضطراب وراثي جسدي متنحي نادر يتميز بتقرن الجلد في راحتي اليد وأخمص القدمين وبالتهاب عدواني تدريجي متقدم يؤدي إلى فقدان الأسنان اللبنية والدائمة. إن إعادة تأهيل الفم بعمل تركيبات ثابتة فوق الزرعات السننية يعتبر ذو ميزات عالية من حيث النواحي الوظيفية والجمالية وأكثر إرضاء للمريض من الأطقم الكاملة المتحركة. في هذا التقرير نفيد نستعرض حالة مريضة مصابة ب (بي أل أس) تبلغ من العمر 19 عاما مرت بعملية خلع كامل لأسنانها ثم أعيد تأهيل فمها بعمل تركيبات ثابتة فوق الزرعات السننية. أظهرت المتابعة بعد عام من العلاج استقرار الحالة من حيث الأداء الوظيفي والناحية الجمالية من دون ظهور أي علامات للتهاب أو فقدان للعظم.

Papillon-Lefevre syndrome (PLS) is a rare autosomal recessive disorder characterized by diffuse palmoplantar keratoderma and precocious aggressively progressing periodontitis, leading to the premature loss of deciduous and permanent teeth. Full mouth rehabilitation with implant supported fixed prosthesis provides numerous advantages over the conventional denture in terms of esthetics, function, longevity of the prosthesis, and patient satisfaction. We report a case of PLS in a 19-year-old female who underwent extraction of all the teeth followed by full mouth rehabilitation with implant supported fixed prostheses. A one-year follow up of the case showed functionally and esthetically stable dental implant with no signs of infection or bone loss.

*Saudi Med J 2013; Vol. 34 (4): 424-427*

*From the Department of Prosthetic Dental Science, College of Dentistry, King Saud University, Riyadh, Kingdom of Saudi Arabia.*

*Received 26th December 2012. Accepted 10th March 2013.*

*Address correspondence and reprint request to: Dr. Abdullah Al Farraj AlDosari, Director of Dental Implant and Osseointegration Research Chair, Department of Prosthetic Dental Science, College of Dentistry, King Saud University, PO Box 60169, Riyadh 11545, Kingdom of Saudi Arabia. Tel. +966 (1) 4677226. Fax. +966 (1) 4678548. E-mail: aalfarraj@hotmail.com*

Papillon-Lefevre syndrome (PLS) is a very rare autosomal recessive disorder characterized by palmoplantar hyperkeratosis and severe early onset periodontitis, affecting the primary and permanent dentition. The prevalence of PLS has been reported as 1-4 per million.<sup>1</sup> The etiopathogenesis of the syndrome is relatively obscure and immunologic, genetic, or possible bacterial etiologies have been proposed.<sup>2</sup> Patients with PLS often present with severe gingival inflammation and periodontal destruction soon after the eruption of primary teeth, leading to premature loss of the deciduous dentition. Due to the vast degree of periodontal breakdown involved at such an early age, the dentist is often the first to diagnose the syndrome. Conventional periodontal therapy generally fails in patients with PLS, and rapid progression of periodontitis frequently results in a severe alveolar bone loss.<sup>1</sup> 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.<sup>3</sup> 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.<sup>4</sup> Conventional periodontal treatment usually fails in PLS patients, resulting in the rapid progression of periodontitis and destruction of alveolar bone. The treatment approach in such cases is extraction and conventional prosthodontic rehabilitation. Recently, endosseous dental implant supported prostheses have been considered as an alternative. Implant-supported prostheses enhance the support, stability, and retention of prostheses. Only a few cases are available in the literature describing the successful use of implants in PLS cases.<sup>5-8</sup> 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. In the present study, a 19-year-old female diagnosed with PLS who underwent extraction of all the teeth followed by prosthetic rehabilitation with implant-supported

porcelain-fused-to-metal restorations, is presented. The case is presented to highlight the advantages of implant supported prosthesis for managing PLS cases.

**Case Report.** A 19-year-old Saudi female patient diagnosed with PLS was referred to the prosthodontist for expert opinion and management of the case. She reported with grade III mobility of all teeth except the second molars. She was diagnosed with severe periodontitis with generalized bone loss in the upper and lower jaw (Figures 1 & 2). Considering her age and the prognosis of the teeth, a decision was made to extract all the teeth except for the second molars of both upper and lower arches. To restore function and esthetics, implant-supported fixed prostheses was considered as the treatment option. During here second visit to the clinic, scaling and root planning was performed for the second molars. All the teeth in the lower jaw except the second molars and the erupting 3rd molar were extracted. After soft tissue relining and occlusal adjustments, the lower immediate transitional removable partial denture was inserted. Two months after extraction of the lower teeth, 6 dental implants (4 x 9 mm, OsseoSpeed™, Astratech Dental Implant System, Mölndal, Sweden) in the canine, first premolar, and first molar region were placed in the lower arch 2 months after the extraction. Bone grafting (Bio-Oss®, Geistlich AG, Switzerland) was carried out in the lower right first premolar and molar region. In the canine as well as premolar region, implants of size 3.5mm diameter and 13 mm length (OsseoSpeed™, Astratech Dental Implant System, Mölnda l, Sweden) were used.

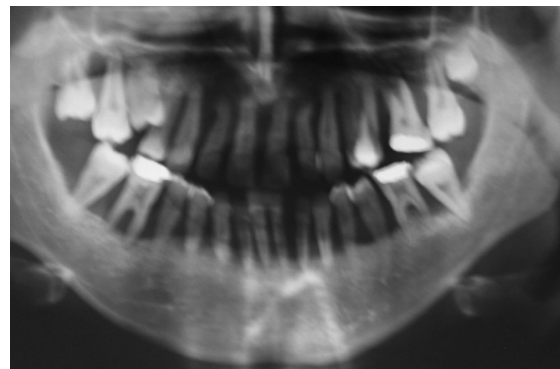
Two months after the extraction of the upper teeth, 8 implants were placed in the central incisor, canine, second premolar, and first molar region on both the sides. Implants (OsseoSpeed™, Astratech Dental Implant System, Mölndal, Sweden) of size 5.0 mm diameter, 9 mm length, and 3.5mm diameter, 11mm length were used. Wider and shorter implants were used at the molar sites. Bone grafting (Bio-Oss®, Geistlich AG, Switzerland, 0.5g, 0.25mm – 1mm) was performed in the premolar and molar region.

After 4 months, all the implants were surgically exposed and impression procedures, and jaw relation recording was carried out (Figure 3). The transitional denture was converted to an interim fixed partial

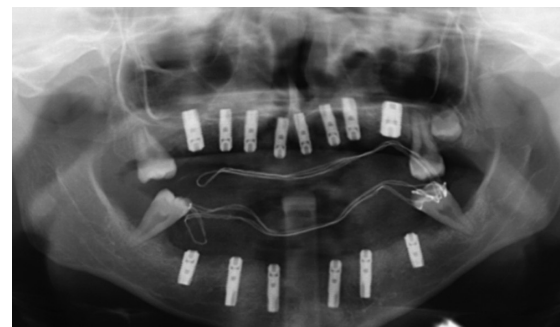
**Disclosure.** Authors have no conflict of interests, and the work was not supported or funded by any drug company.



**Figure 1** - Intraoral photograph of the patient showing inflamed gingiva, mobility teeth and spacing.



**Figure 2** - Pre-treatment panoramic radiograph showing generalized bone destruction.



**Figure 3** - Panoramic radiograph, after the placement of implants in upper and lower jaw.

denture. A laboratory putty index (Aquasil DECA®, Dentsply Caulk, Milford, DE, USA) of the established tooth position was made. The prosthesis was inserted and the vertical dimension, esthetics, phonetics, and occlusion were evaluated (Figure 4).

The framework was then sent to the laboratory for porcelain application. Finally, esthetic and occlusion corrections were made on the prosthesis. The occlusion of the prosthesis was adjusted to achieve simultaneous



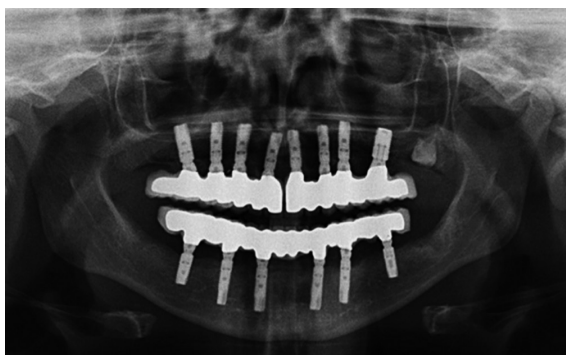
**Figure 4** - Interim Fixed Partial denture in occlusion.



**Figure 5** - Post insertion of the final prosthesis.



**Figure 6** - Intraoral photograph of the restored dentition after one year.



**Figure 7** - Post-operative radiograph after one year.

centric relation contact and canine protected occlusion. In the final visit, the abutments were torqued to 25 Ncm<sup>2</sup> and the screw holes were sealed with light polymerizing provisional resin (Fermit-N; Ivoclar Vivadent, GmbH, Bolzano, Italy) and composite resin (Tetric Ceram HB; Ivoclar, Vivadent, GmbH, Bolzano, Italy). The final prosthesis was cemented using temporary cement (TempBond NE, Kerr/Sybron, Romulus, MI, USA). After the final insertion of the prosthesis, the interfaces were checked for accuracy radiographically. The occlusal vertical dimension, esthetics, phonetics, occlusion, and patient satisfaction were evaluated (Figure 5). Post insertion instructions regarding maintenance of oral hygiene, use of water jet (Waterpik® Ultra Cordless Dental Water Jet, Surrey, UK) and dental floss were provided.

Periodic follow up of the case was carried out up to one year (Figures 6 & 7). Oral hygiene maintenance and occlusion was evaluated during the recall visits. She was satisfied with the aesthetics and functioning of the prosthesis.

**Discussion.** Papillon-Lefèvre Syndrome is a devastating disease process characterized by rapid destruction of the dental alveolar complex. Early extraction of permanent dentition and prosthodontic rehabilitation has been suggested as a method of managing such cases.<sup>5,6</sup> Our patient had poor prognosis as most of the teeth had grade III mobility. The second molars were retained to be used as abutments, and also to maintain the vertical dimension of occlusion.

So far only 4 cases are available in the literature, which described dental implant supported prosthetic rehabilitation in patients with PLS. Implants will act as an ankylosed tooth if placed before the growth of the alveolar process has stopped, and thus a contraindication in young children.<sup>9</sup> Prosthetic rehabilitation becomes difficult in patients with atrophic mandibles and maxillae. This may call for various additional surgical techniques such as distraction osteogenesis, bone augmentation procedures, and nerve lateralization to achieve adequate bone level in atrophic jaws before implant placement. Some studies have reported only implant placement for severely atrophic mandibles as the surgical procedures could lead to possible bone fracture.<sup>10</sup> Normal healing was observed post-surgery. After one year of follow-up, the clinical and radiological conditions of the osseointegrated implants and the denture were assessed and no signs of infection or unexpected bone loss around the implants were noticed.

In conclusion, the case presented highlights the use of dental implants in the successful rehabilitation of

PLS patients and to provide excellent functional and esthetic dental rehabilitation.

**Acknowledgment.** *The author would like to acknowledge Dr. Saleh Al Bazie for his participation in the surgical part of the treatment.*

## References

1. Shapira J, Eidelman E, Fuks A, Hacham-Zadeh S. Treatment of Papillon-Lefevre syndrome with chemotherapy: report of cases. *Spec Care Dentist* 1985; 5: 71-74.
2. Dalgic B, Bukulmez A, Sari S. Eponym: Papillon-Lefevre syndrome. *Eur J Pediatr* 2011; 170: 689-691.
3. 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.
4. Pilger U, Hennies HC, Truschneegg A, Aberer E. Late-onset Papillon-Lefevre syndrome without alteration of the cathepsin C gene. *J Am Acad Dermatol* 2003; 49 (5 Suppl): S240-S243.
5. 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.
6. 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.
7. Senel FC, Altintas NY, Bagis B, Cankaya M, Pampu AA, Satiroglu I, et al. A 3-year follow-up of the rehabilitation of Papillon-Lefevre syndrome by dental implants. *J Oral Maxillofac Surg* 2012; 70: 163-167.
8. Ahmadian L, Monzavi A, Arbabi R, Hashemi HM. Full-mouth rehabilitation of an edentulous patient with Papillon-Lefevre syndrome using dental implants: a clinical report. *J Prosthodont* 2011; 20: 643-648.
9. Oesterle LJ, Cronin RJ, Jr., Ranly DM. Maxillary implants and the growing patient. *Int J Oral Maxillofac Implants* 1993; 8: 377-387.
10. Etoz OA, Ulu M, Kesim B. Treatment of patient with Papillon-Lefevre syndrome with short dental implants: a case report. *Implant Dent* 2010; 19: 394-399.

## Illustrations, Figures, Photographs

Four copies of all figures or photographs should be included with the submitted manuscript. Figures submitted electronically should be in JPEG or TIFF format with a 300 dpi minimum resolution and in grayscale or CMYK (not RGB). Printed submissions should be on high-contrast glossy paper, and must be unmounted and untrimmed, with a preferred size between 4 x 5 inches and 5 x 7 inches (10 x 13 cm and 13 x 18 cm). The figure number, name of first author and an arrow indicating "top" should be typed on a gummed label and affixed to the back of each illustration. If arrows are used these should appear in a different color to the background color. Titles and detailed explanations belong in the legends, which should be submitted on a separate sheet, and not on the illustrations themselves. Written informed consent for publication must accompany any photograph in which the subject can be identified. Written copyright permission, from the publishers, must accompany any illustration that has been previously published. Photographs will be accepted at the discretion of the Editorial Board.