

Combined orthodontic and periodontic treatment in a child with Papillon Lefèvre syndrome

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ABSTRACT

نستعرض في هذا المقال حالة طفلة تبلغ من العمر 9 أعوام ومصابة بمتلازمة بابيلون لوفيفر، وقد تم تقويم أسنانها وذلك بعد مرور 24 شهراً من بدء المضادات الحيوية بالإضافة إلى علاج اللثة كل 6 أسابيع. لقد قمنا ببدء علاج تقويم الأسنان وذلك بعد استقرار حالة اللثة لديها وذلك من أجل تصحيح وضعية الأسنان، وجانب الوجه، وحماية الفك العلوي. وبعد العلاج اللثوي وتقويم الأسنان وبسبب فشل اكتشاف المشعشة المصاحبة للورم الفطري في جوف الأسنان فقد قمنا باللجوء إلى تقويم الأسنان الثابت بالإضافة إلى إتاحة المساحة المناسبة لظهور الأسنان الدائمة. لقد وجدنا أن الجمع بين علاج اللثة وتقويم الأسنان للمرضى المصابين بمتلازمة بابيلون لوفيفر قد يكون ناجحاً بعد تعاون التخصصات اللازمة لعلاج مثل هذه الحالات بالإضافة إلى المتابعة الدقيقة والمستمرة. لقد قمنا باستعراض حالة مريضة مصابة بمتلازمة بابيلون لوفيفر تمت متابعتها على مدى عامين، وقد قمنا باتباع علاج اللثة وتقويم الأسنان، بالإضافة إلى استشارة المعالجة السنوية التعويضية التي اعتمدت خطة العلاج المتبعة.

A 9-year-old girl with Papillon-Lefèvre syndrome (PLS) was treated orthodontically 24 months after the start of mechanical and antibiotic therapy in adjunct with periodontal treatment every 6 weeks. After achieving stable periodontal conditions, orthodontic treatment was commenced to correct the teeth position, facial profile, and maxillary protraction. Following the combination therapy and a failure to detect *Actinobacillus actinomycetemcomitans* from any site in the oral cavity, orthodontic treatment with a fixed appliance was performed aside from creating space for eruption of permanent teeth. We found that combined periodontal and orthodontic treatment of PLS may be successful with a complex interdisciplinary regimen and close follow up. This is a 2-year follow-up case report of a girl with PLS. Orthodontic and periodontic therapy were offered using combined treatments of orthodontic and periodontal with the benefit of prosthodontic consultation, resulting in a treatment plan.

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Papillon-Lefèvre syndrome (PLS) is described as a rare autosomal recessive trait affecting between one and 4 people per million, irrespective of gender and race. The syndrome, characterized by palmo-plantar keratosis, manifests in the oral cavity as a form of early onset aggressive periodontitis in both the deciduous and permanent dentition.¹ Before primary dentition, the gingival and mucosal surfaces appear normal; however, in post-eruption there is a rapid periodontal destruction with exfoliation of the deciduous dentition by 4-5 years of age in most cases.² At the age of 4, prepubertal periodontitis with considerable inflammation develops and proceeds until all deciduous teeth are lost. The inflammation disappears during the edentulous period, but the process repeats itself when permanent dentition erupts.³ Most teeth are lost during the early teen years, although late-erupting wisdom teeth have often been spared. Severe destruction of the alveolar bone often occurs, leaving atrophic jaws. Microbiologic monitoring and clinical examination following various treatment modalities has highlighted a close relationship between the presence of *Actinobacillus actinomycetemcomitans* and periodontal destruction in patients with PLS.⁴ Furthermore, current literature suggests that the success of periodontal treatment in patients with PLS is highly variable and often leads to partial or complete tooth loss.⁴ Previous study⁵ reported patients with PLS have the characteristics of skeletal Class III malocclusion. Therefore, our aim was to establish first the Class I malocclusion to enhance the child's facial skeletal

appearance, and then maximize the future prosthetic appliance retention that may be needed.

Case Report. This report describes the outcome of an interdisciplinary treatment modality for a child with PLS and the subsequent 24-month follow-up. In February 2011, a 9-year-old girl was referred from a private clinic to the Periodontal Department, College of Dentistry, King Saud University, Riyadh, Saudi Arabia for evaluation of her missing teeth. She was diagnosed with PLS by her dermatologist at the age of 5 (Figures 1A and 1B). Her medical history, as reported by the dermatologist, indicated hyperkeratotic lesions on the palms, knuckles, knees, elbows, and soles. She was the sixth of eight children, and her eldest sister and younger brother exhibited the same condition. The children were products of a consanguineous marriage free from PLS and other cutaneous disorders.

Several treatment regimens have been recommended in the literature; however, a definitive treatment protocol has not been established. This case report evaluates the success of combined therapy in managing a patient with PLS.

Periodontal examination. She was referred with a chief complaint of “backward positioning of upper front teeth.” Her dental history revealed premature exfoliation of primary teeth at the age of 4 years. The permanent upper central incisors, first molars, and all the lower incisors were erupting with acute gingival inflammation, and an initial signs of necrosis around the first molars. In addition, there was a severe loss of bone height buccolingually and apicocoronally and subsequent loss of facial profile around the first permanent molars. The oral mucosa was normal in color and consistency. Subgingival plaque samples were taken for microbiological investigation at 3-month intervals using sterile paper points inserted into the pockets following removal of supragingival plaque. They were immediately placed into vials containing 1 ml anaerobically sterilized half-strength Ringer’s solution. This solution was sonicated for 10 seconds after which a sterile cotton swab was used to spread it on freshly prepared trypticase-soy agar plates with serum, bacitracin, and vancomycin for selective cultivation of *Actinobacillus actinomycetemcomitans*. After 3-5 days incubation at 36°C in air with 5% carbon dioxide, standard suspensions of *Porphyromonas*

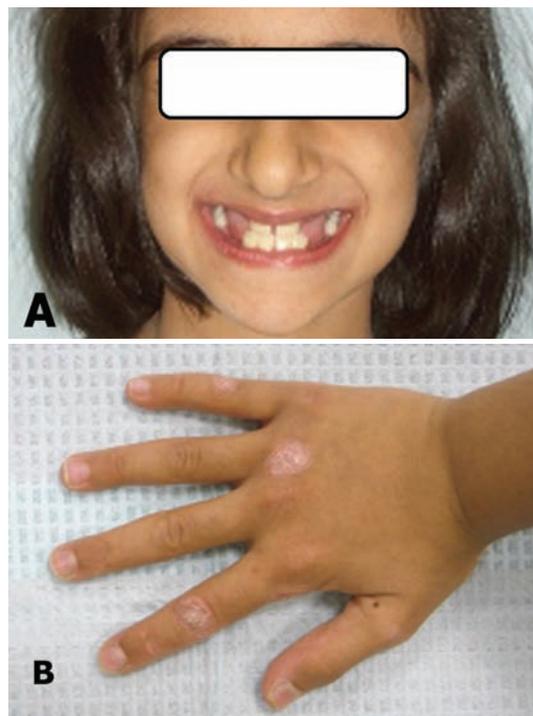


Figure 1 - A photograph showing the A) facial view and B) hand view of patient diagnosed with Papillon-Lefèvre syndrome.

gingivalis, *Prevotella intermedia*, *Actinobacillus actinomycetemcomitans*, and *Fusobacterium nucleatum* were also fixed to the wells. The test was positive for *Actinobacillus actinomycetemcomitans* with a small amount of supragingival plaque and abundant subgingival plaque.

Orthodontic examination. Her dental history revealed that all primary teeth were exfoliated prematurely at the age of 4 years, orthodontic records (namely, orthodontic study models, panoramic radiograph, cephalometric radiograph, hand and wrist radiograph, and extra- and intra-oral photos) were taken for further analysis. Examination of the records highlighted the maxillary deficiency with retroclination of the upper incisors. Extra-oral examination showed an ovoid facial profile with a concave lateral profile and competent lips (Figure 2A). Intra-oral examination revealed a reverse overjet (-2 mm) and 20% incisal vertical overlap (overbite). The maxillary arch contained the permanent central incisors and first molars, while the mandibular arch contained the permanent central and lateral incisors and first molars. Right and left molars showed a Class III relationship. Panoramic radiograph analysis showed severe horizontal resorption of alveolar bone which gave the teeth a “floating-in-air” appearance (Figure 2B). Cephalometric analysis showed

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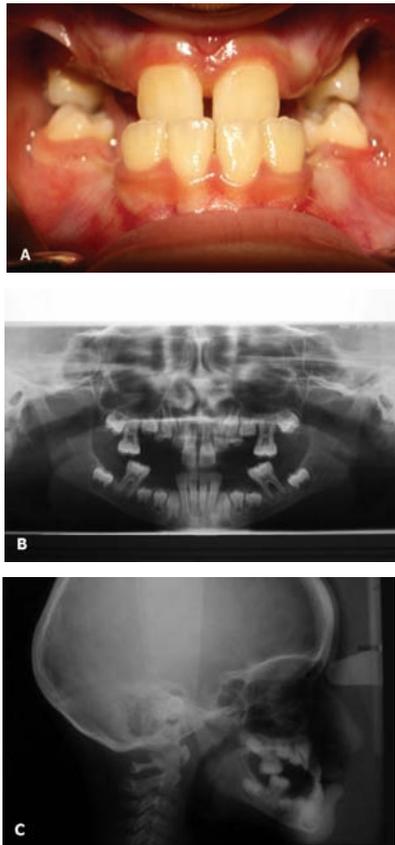


Figure 2 - A photograph showing the A) pre-treatment oral view, B) panoramic radiograph analysis showing the pre-treatment, and C) the x-ray showing the pre-treatment lateral view

the maxillary growth deficiency in the anterioposterior dimension (sella, nasion, A point [SNA]= 79°) with retroclination of the upper incisors (maximum incisor to nasion A point [U1-NA] = 14°/0mm; **Figure 2C**). The lower facial height measured 51%, and the Frankfort to mandibular plane angle measured 20°, indicating a mesiocephalic facial type. The inter incisal angle was greater than the normal (155°) revealing an upright position of the upper and lower incisors. Both the upper and lower incisors were in the retruded position. Additionally, soft tissue analysis showed a retruded upper lip (H line = -6 mm).

Periodontal treatment. Anti-infective therapy comprising of oral hygiene instructions, supragingival, and subgingival instrumentation with irrigation using 0.12% chlorhexidine mouth rinse was started as the initial regime. Also, systemic antibiotics of amoxicillin 250 mg (twice/day) and metronidazole (250 mg 3/day) was administrated for 2 weeks. During the orthodontic phase of the treatment, she was kept in a maintenance program, which included oral hygiene instructions



Figure 3 - A photograph showing orthodontic treatment by fitting orthodontic bands with triple tubes (Roth 0.022-inch; West Collins Orange, CA USA) on the maxillary first permanent molars.

and oral prophylaxis once every 6 weeks to 2 months, depending on the oral hygiene status of the patient.

Orthodontic treatment. Dental treatment was started once all the permanent first molars erupted and were sealed with fissure sealants. She was kept under a strict periodontal maintenance regimen. She was assessed at 2- to 4-week intervals, and when severe inflammation of the gingiva, indicated by red coloring, sponge-like consistency, and spontaneous bleeding was observed, an antibiotic regimen was administrated to control the inflammation.

Orthodontic treatment was started by fitting orthodontic bands with triple tubes (Roth 0.022-inch; West Collins Orange, CA USA) on the maxillary first permanent molars (**Figure 3**). An alginate impression of the maxillary arch was taken, and the bands were seated in the impression and sent for fabrication of a Nance button space maintainer with hooks to accommodate a protraction face mask. The hooks were extended from the molar bands to the distal surface of the maxillary canines. Additionally, the same orthodontic bands with double tubes were fitted to the mandibular first molars, and the same process was repeated. A mandibular lingual arch space maintainer with adjustment loops mesial to the molars was fabricated. Glass Ionomer cement (3M-Unitek, Monrovia, CA, USA) was used to cement both the Nance button space maintainer and mandibular lingual arch space maintainer. An AD® Protraction face mask (West Collins Orange, CA USA), used for maxillary advancement, was adjusted to the patient's forehead and chin. Supplementary oral rubber elastics (5/16") (Orthotechnology, Inc., Tampa, FL, USA) were fitted to the face mask hooks and to the horizontal elastic bar on the face mask (**Figure 4**). A light continuous force was used during the entire treatment (150g force per side) for 12 months. She was instructed to wear the face mask for at least 14 hours daily as recommended.⁶ A recent review by Yepes et al⁷ revealed that there is no scientific evidence that allows definition of the force magnitude when

using the maxillary protraction face mask treatment in Class III patients.⁷ However, the force magnitude values reported in the literature range between 180 and 800 g per side as during growth modification, tooth movement is undesirable and the objective is to correct jaw discrepancy not to move the teeth to camouflage it.⁷ In this case, we chose a lesser force in consideration of the periodontal condition of the maxillary molars to prevent further inflammation and destruction of the periodontal and bony supporting tissues surrounding the maxillary permanent molar.

Following 3 months of maxillary protraction, the maxillary central incisors were bonded with 0.022" brackets (3M-Unitek, Monrovia, CA, USA) using Transbond® adhesive (3M-Unitek). A 2 X 2 utility arch wire (0.016" x 0.016" Blue Elgiloy; Rocky Mountains Inc., Denver, CO, USA) was used for proclination of the upper incisors. At the end of 12 months, the maxillary Nance button space maintainer and brackets were removed. Final orthodontic observations were recorded, and an upper Haweley retainer was fitted to the upper arch. The mandibular lingual arch space maintainer was used until all the posterior permanent teeth erupted. She was instructed to wear the appliance full time and was followed up frequently. At one, 3, 6, and 12 months, the wraparound retainer was adjusted and the mandibular lingual arch space maintainer was checked. Also, infection and bacterial plaque were controlled by means of mechanical and chemical methods during those visits. At the 12-month follow-up visit, extra- and

intra-oral photographs were taken to document the stability of orthodontic results (Figure 5).

Figure 6A shows the results of combined periodontal and orthodontic treatment at age 12, after a retention period of 2 years. A 3-mm maxillary advancement was achieved, as was proclination of the upper incisors by 1.5 mm. An overjet of 1.5 mm with a 30% overbite was recorded, and coincident maxillary and mandibular



Figure 5 - A photograph showing the orthodontic results during the 12-month follow-up.

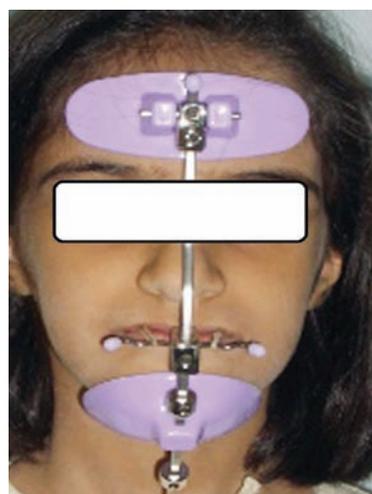


Figure 4 - A photograph showing of an AD® protraction face mask (West Collins Orange, CA USA) for maxillary advancement to the patient's forehead and chin. Supplementary oral rubber elastics (5/16") (Orthotechnology, Inc., Tampa, FL, USA) were fitted to the face mask hooks and to the horizontal elastic bar on the face mask.

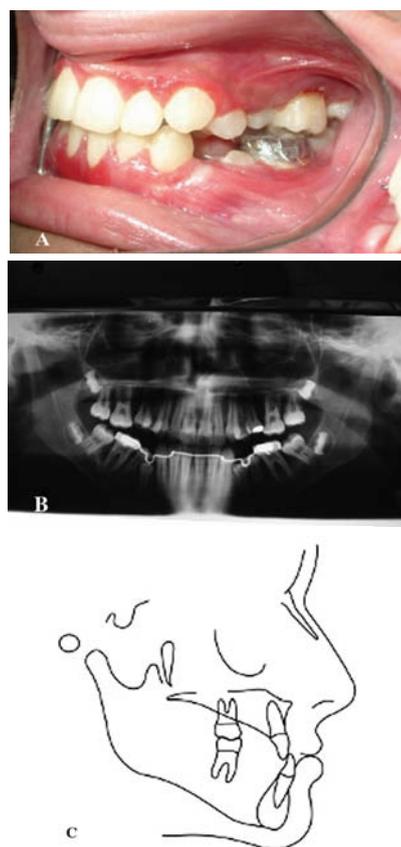


Figure 6 - Cephalometric analysis after 2-year follow up A) the results of combined periodontal and orthodontic treatment at age 12, after a retention period of 2 years. B) X-ray showing the insufficient space for canines and premolar in the upper and lower dental arches. C) Sketch of the results after the 2-years follow up.



Figure 7 - A photograph showing the results of combined periodontal and orthodontic treatment at age 12, after a retention period of 2 years.

midlines were achieved. The panoramic radiograph highlights severe horizontal bone loss around all first molars and moderate bone loss at all incisors (Figure 6B). Space analysis showed insufficient space for canines and premolar in the upper and lower dental arches (Figure 6C). However, long-term prognosis of the first permanent molars in both dental arches remains critical (Figures 6C and 7).

Discussion. Papillon-Lefèvre syndrome is manifested early, usually before 4 years of age. The time and sequence of eruption of the deciduous and permanent teeth are normal, although both are lost prematurely due to severe periodontal disease.⁴ The periodontitis associated with PLS is usually difficult to control. Effective treatment includes the extraction of the primary teeth combined with oral antibiotics and professional teeth cleaning.^{8,9} It is reported that tretinoin (TEGISON®, Hoffmann-La Roche, USA) and acitretin (TEGISON®, Hoffmann-La Roche, USA) modulate the course of periodontitis and preserve the teeth.¹⁰ Orthodontic correction is a documented approach for early treatment of patients with mild Class III malocclusion.^{11,12} In normal situation for orthodontic treatment of Class III, periodontal tissue should be in optimal health condition. Patients with affected periodontal tissues are at high risk of losing their dentition.¹³ In the literature, 4 cases were revealed that orthodontic treatment combined with an antibiotic regimen, if well-planned and followed, can successfully lead to the maintenance of healthy dentition.¹⁴⁻¹⁷ Therefore, mild skeletal Class III malocclusion in patients with PLS can be corrected utilizing early facemask therapy.

In our case, space analysis showed insufficient space for canines and premolars in the upper and lower dental arches. However, long-term prognosis of the first permanent molars in both dental arches remains critical. However, it should be mentioned that extraction of the first permanent molars probably would likely result in the best long-term stability of orthodontic alignment of the canines and premolars. Early treatment of Class III malocclusion is recommended for PLS patients to achieve normal maxillary growth, to prevent traumatic occlusion of the anterior teeth, and to improve the patient's facial profile and self-esteem.

In conclusion, this case report demonstrates that individually, with successful combined mechanical and antibiotic periodontal therapy of PLS periodontitis, moderate orthodontic tooth movements may be possible with a complex interdisciplinary treatment regimen. Also, the effectiveness of early face mask treatment for PLS patients can be achieved with optimal periodontal therapy.

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