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## PAPILLON-LEFÈVRE SYNDROME: ORAL ASPECTS AND TREATMENT

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**Abstract**

Papillon-Lefèvre syndrome (PLS) is a rare disorder characterized by diffuse palmoplantar erythematous, fissured hyperkeratosis and aggressive periodontal disease that starts in the early periods of childhood. Periodontal disease occurs with the early loss of deciduous teeth at the age of 2 to 4 years, followed by the loss of permanent teeth during adolescence. Prosthodontics management of PLS patients is very complex and sometimes requires invasive therapeutic treatments. Early diagnosis is essential for correct treatment management avoiding the possibility that patients are early edentulous. Management could be a conventional periodontal treatment and pharmacological therapy but in severe cases, digital techniques, could be help the clinician for increased patient comfort and minimized tissue damage.

**Key words:** Papillon-Lefèvre syndrome, Treatment, Management

## **Introduction**

Papillon-Lefèvre syndrome (PLS) is an autosomal recessive trait characterized by the presence of palmo-plantar hyperkeratosis and premature loss of the deciduous and permanent teeth (Haneke; 1979). The periodontal lesions appear early after the start of the deciduous dental elements in the arch (Dekker& Jansen, 1958). Periodontal tissues are inflamed with evident signs of bleeding of the gums, presence of periodontal pockets, tooth mobility, loosening without signs of root resorption and anomalies in the order of their eruption. The same clinical signs occur for permanent dental elements shortly after their appearance in the arch. Other clinical signs than those described in Papillon-Lefèvre syndrome are recurrent pyogenic skin infections, intra-cranial calcifications and mental retard (Sachdeva, Kalra& Kapoor, 2012).

## **Genetic**

PLS (PPK with periodontitis, MIM 245000) is an autosomal recessive disorder caused by mutations in the CTS gene, coding for the proteolytic enzyme cathepsin C, a lysosomal cysteine protease required for the activation of granule-associated serine proteases in immune - inflammatory cells (Castori et al., 2009). Cathepsin C is composed of 3 polypeptidic chains

joined by non-covalent interactions and more than 60 CTSC mutations capable of altering the folding and function of proteins have been identified. In fact, the expression of the cathepsin C gene is characteristic of epithelial regions such as the keratinized oral gingiva in patients with PLS.

### **Oral clinical aspects**

Patients with Papillon-Lefèvre syndrome presents first clinical signs in the oral cavity as soon as the deciduous teeth erupt. Periodontal manifestations become apparent by the age of 2-3 years. As is evident from literature (Dhanrajani, 2009) the clinical manifestations may be different.

The oral manifestations of primary teeth are represented by plaque accumulation, severe gingivitis, periodontitis and multiple caries.

Once the eruption of the deciduous teeth has been completed, erythematous oral mucosa with perioral lymphadenopathies and halitosis can be appreciated. Normally, no sequence, timing, shape and structure anomalies are observed, although in some cases may have microdontia and incomplete root formation (Fahmy, 1987). The rapid progress of inflammatory index and the progressive of periodontal disease is manifested with redness and swelling in the gingiva in association at periodontal pockets and severe bone resorption. Often gingival tissues near the dental elements are inflamed, edematous, tender to palpation and periodontal abscesses are noticed on teeth, while those of edentulous regions appeared healthy. Generally follows premature exfoliation of all deciduous teeth by the age of 4 to 5.

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After patients will loss of all deciduous teeth, gingival inflammation disappears, and the oral mucosa takes on a healthy appearance (Ashri, 2008). Usually level of patient's cooperation is low (Abou Chefin et al., 2019).

Oral aspects in permanent dentition are similar to those of deciduous dentition. The time of eruption of permanent teeth is physiological in timing and development and shape were also normal. When teeth erupt the gingiva becomes red and edematous and after their eruption it becomes edematous and bleeding. Characterized by severe gingival inflammation, with high bleeding indexes, associated with plaque accumulation, deep periodontal pockets begin to form. Periodontal defects are severe and presence of gingival abscesses, purulent exudates and offensive halitosis are characteristic. Dental elements present high mobility, fluttering of the anterior group and loss of the vertical dimension. Spontaneous losses of the dental elements frequently occur and gingival abscesses associated with migration of teeth causing malocclusion. For this reason patient's presented a reduce facial height and senile appearance. Radiographic investigations show generalized alveolar bone loss and migration of teeth whit no evidence of root resorption. Associated periodontal support loss and an advanced state of bone resorption are characteristic in Papillon-Lefèvre syndrome

### **Treatment**

Treatment in deciduous teeth, if an early diagnosis is made and early treatment is performed could be non-surgical and involves a conventional periodontal treatment in form of scaling and root planning. In association with periodontal antibiotic amoxicillin and metronidazole (250 mg, 3 times daily) for one week along with a mouth rinse (0.2% chlorhexidine gluconate, 10

mL twice daily) is prescribed to the patient. It is important to educate the patient on correct oral hygiene and organize a follow-up visit every month for scaling and checking the condition of the patient. This therapeutic approach, if performed late or ineffective, will necessarily lead to extract the dental elements (Ullbro et al., 2003). When deciduous teeth are lost, prosthetic therapy will be necessary to replace the lost elements. The prosthetic treatment will be age-specific and will be made with partial or complete dentures (AIBarrak et al., 2016). During mixed dentition period, lost teeth are replaced with removable prostheses, restoring lost function and aesthetics. Patients will be checked regularly until the exchange with permanent teeth and prostheses will be modified during growth.

The treatment of permanent teeth is more complex but may be similar to that just described for deciduous teeth if early intervention is performed. Local debridement, conventional periodontal treatment and systemic antibiotics can be used in combination and improvement in the inflammatory state is possible. This approach can be considered above all in relation to age and psycho-social impact (Jain, Gupta & Prakash, 2005). Non-surgical therapy needs to educate patients on proper oral hygiene and schedule for a follow-up visit every month for scaling treatment and checking periodontal condition (Ullbro, Brown & Twetman, 2005).

When tooth mobility becomes high and periodontal abscesses were recurrent, all teeth must be extracted.

First therapeutic option to restore lost teeth is by means of complete mobile prostheses on both arches. This therapeutic option allows the restoration of function and aesthetics quickly but causes some discomforts such as taking impressions with pastes (Lampraki et al., 2016; Lee &

Park, 2009). It is known that the presence of mobile teeth complicates the dental impression process and can result in accidental tooth extraction.

For this reason some authors (Millet et al., 2019) suggest the possibility of using new digital techniques to simplify procedures. The introduction of digital workflows in dentistry allows to extract the teeth and insert the complete dentures in the same session reducing the psychological trauma of edentulism. By acquiring intraoral scanners of both arch, before performing the extractions, it is possible to fabricate the prostheses with CAD-CAM technique. These will be inserted and adapted intraorally after extracting the teeth. Digital approach increased patient comfort, reduces the work of the clinician and laboratory but has limitations in the acquisition of some areas of soft tissue. It is also known that complete inferior prosthesis may be unstable, especially in the lower arch (Goodacre, Goodacre & Baba, 2018).

The reduced stability of the lower total prostheses is a known problem in dentistry but the use of osseointegrated implants in children or adolescent is a topic much discussed in dentistry (Oesterle, Cronin & Ranly, 1993; Brugnolo et al., 1996) even if today there are many scientific works that encourage their use (Giannetti et al., 2010).

To solve this problem, some authors have proposed (Woo et al., 2003) the possibility of stabilizing the lower prosthesis by insert implants. The placement of implants stabilizes the prosthesis and, as shown by this case report, from the follow-up radiographs, preserves alveolar bone and shows successful osseointegration.

## **Conclusion**

Papillon-Lefèvre syndrome occurs at an early age, generally before the age of four. A multidisciplinary approach is very important, dermatologist and dentist collaborate in the care of patients with PLS. Aggressive periodontitis affecting both teeth, deciduous and permanent, must be managed quickly by trying to procrastinate the loss of teeth as much as possible. However, tooth loss will be inevitable and therapy must be managed correctly. The therapy will be different and will have to take account the patient's psychological state, age and degree of tooth mobility.

Dental knowledge and techniques are now very advanced and allow to limit patients' discomfort.

In conclusion, the management of periodontal treatment of patients with PLS remains complex. The new knowledge and new materials available today in periodontology induce to be optimistic even if dental management is still under study.

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