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Opinion Article

Early-Onset Periodontal Breakdown in Papillon-Lefèvre Syndrome: Pediatric Insights

Kamilla Rosen*

Department of Clinical Dentistry, University of Bergen, Bergen, Norway

DESCRIPTION

Papillon-Lefèvre syndrome is an uncommon inherited disorder that typically appears in early childhood and affects both the skin and oral cavity. In dental studies, it is particularly significant because of the rapid and severe periodontal destruction that occurs soon after the eruption of primary teeth. The condition follows an autosomal recessive inheritance pattern and is linked to changes in genes that regulate immune cell function, especially those involved in the activity of neutrophils. These biological changes result in an impaired defense mechanism within periodontal tissues, allowing bacterial populations to trigger aggressive inflammatory reactions that the body is unable to regulate effectively.

The earliest oral signs usually become apparent shortly after the eruption of the primary dentition. Parents may observe swollen gums, persistent bleeding during routine brushing, and increasing mobility of teeth that have only recently erupted. Despite standard oral hygiene practices, inflammation does not resolve and instead progresses in severity. Within a relatively short period, destruction of the periodontal ligament and surrounding alveolar bone occurs, leading to premature exfoliation of primary teeth. This pattern of destruction is often repeated when permanent teeth erupt, producing a similar sequence of inflammation, attachment loss, and eventual tooth loss at an early age.

The dermatological component of the condition is typically characterized by thickened, scaly skin on the palms and soles. This hyperkeratotic presentation may vary in intensity among affected individuals but is generally persistent over time. The coexistence of skin and oral manifestations provides important diagnostic clues, especially when both features are observed in a young child. However, the severity of oral involvement often exceeds that of the skin changes, making dental symptoms the primary concern in clinical management.

At the biological level, the condition is associated with alterations in genes that influence lysosomal enzyme activity in immune cells. These changes interfere with the normal ability of neutrophils to eliminate bacterial pathogens effectively. As a result, periodontal tissues become highly susceptible to infection-driven inflammation. The microbial environment within the oral cavity contains bacteria that are normally present in low levels in healthy individuals, but in this condition, they trigger exaggerated tissue responses due to immune dysfunction.

The destruction of periodontal structures in affected children is not solely a consequence of bacterial accumulation but is largely driven by the inability of the host immune system to regulate inflammatory responses. Once inflammation begins, it tends to persist and intensify, leading to progressive breakdown of connective tissue fibers and supporting bone. The speed of this process is unusually rapid compared to typical forms of periodontal disease, which makes early intervention challenging and often less effective in preventing tooth loss.

Diagnosis of Papillon-Lefèvre syndrome is based on clinical findings supported by genetic testing when available. The combination of early-onset severe periodontitis and palmoplantar skin changes strongly suggests the condition. Dental radiographs often reveal rapid bone loss around affected teeth, further supporting the diagnosis. Early identification is important, as it allows for timely preventive strategies aimed at slowing disease progression, even though complete prevention of tooth loss remains difficult.

CONCLUSION

Papillon-Lefèvre syndrome in children represents a complex condition involving genetic immune dysfunction that leads to severe periodontal disease and early tooth loss, accompanied by characteristic skin changes. Its early onset and rapid progression present significant challenges in dental management and child development, requiring early recognition and long-term multidisciplinary care to address both functional and psychosocial consequences. Psychological support is also an important aspect of care. Children affected by early tooth loss

Correspondence to: Kamilla Rosen, Department of Clinical Dentistry, University of Bergen, Bergen, Norway, E-mail: kamillarosen@scm.nw

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may experience reduced self-esteem and difficulties in social integration. Family counseling can help caregivers understand the chronic nature of the condition and the importance of maintaining consistent oral care practices despite limited long-term success in preserving dentition.