

# Syndromes and Periodontal Disease-An Insight

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## ABSTRACT

Periodontal disease is multifactorial in nature but is primarily caused by the bacteria present in dental plaque. Periodontal disease is often seen as oral manifestation of certain syndromes and genetic disorders either as a result of the syndrome or as an exacerbation of an already existing condition. These syndromes result in alterations in the immune system or the structure of periodontal tissues which further enhances the susceptibility of the individual to gingival inflammation, clinical attachment loss and bone resorption. Non-surgical, antimicrobial and surgical periodontal therapy remains the treatment of choice in such patients but to what extent these treatment modalities will be effective in saving the natural teeth is a topic of debate. The purpose of this article is to review the various syndromes associated gingival and periodontal diseases so that it becomes uncomplicated for the clinician to make a differential and final diagnosis with proper management of the case.

**Key words:** Syndrome, Periodontal, Bone loss, Disease, Genetic.

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## INTRODUCTION

A syndrome is a concurrence of a set of signs and symptoms related with a particular disease or disorder [1]. Often the cause of a syndrome is not known; when it becomes associated with a cause it becomes a disease. In this context, periodontal disease is often considered as a syndrome because of its complex etiology [2]. Periodontitis is considered to be a group of pathological conditions leading to attachment loss and bone loss. Also they are a group of polygenic and polymicrobial infections, the clinical expression of which can be altered by environmental factors, systemic diseases and other factors [3]. Aggressive periodontitis was referred as Gottlieb syndrome by some researchers due to the typical incisor and molar involvement [4]. There are certain syndromes associated with periodontal findings that are systemic and genetic in nature. Certain syndromes have a strong association with periodontal disease, however in other syndromes presence of periodontal signs and symptoms can be a mere coincidence. The literature however lacks comprehensive and long term studies on this subject.

## Common syndromes associated with periodontal disease

### Down syndrome

The literature reports 96% of Down's syndrome patients suffer from periodontitis [5]. The general findings include

congenital cardiac anomalies, mongoloid slanting of eyes, flat face and patients prone to acute necrotizing lesions. The periodontal findings include early-onset periodontitis with premature loss of mandibular anterior teeth [6].

### Papillon-lefevre syndrome

The general findings include hyperkeratosis of palms and soles, knees, dorsum of fingers and toes. Male-to-female ratio of incidence is equal. The onset is between the 1st and 5th years of life. The periodontal findings include severe alveolar bone loss, exfoliation of both deciduous and permanent teeth [7].

### Metabolic syndrome

It is characterised by increased central obesity, increased triglyceride levels with reduced concentration of high density lipoprotein cholesterol, abnormal blood pressure and blood sugar levels. There are many reports in the literature suggesting increased prevalence of periodontal disease in patients suffering from metabolic syndrome [8].

### Haim-Munk syndrome

It is similar to Papillon-Lefevre syndrome. Skin manifestations are more severe and periodontal disease milder. There are callous patches of skin on palms and the soles. Periodontal findings include progressive periodontal disease [9].

### Ehlers-Danlos syndrome

The syndrome involves defect in collagen synthesis. The general findings include hypermobility of joints and marked elasticity of skin. The periodontal findings include

gingival bleeding, marked periodontal disease and generalized membranous gingival enlargement [10].

### Lazy leukocyte syndrome

It is characterised by severe neutropenia and the affected individuals are more prone to opportunistic infections due to the immunocompromised state. Periodontitis is one of the oral manifestations of this syndrome [11].

### Leukocyte adhesion deficiency syndrome

This syndrome is characterised by defects in neutrophil chemotaxis and phagocytosis. This leads to enhanced susceptibility to bacterial infections. Aggressive periodontitis is seen in these patients involving both the deciduous and permanent dentition [12].

### Chediak-Higashi syndrome

The findings include involvement of cell organelles of cells like platelets, melanocytes and phagocytes. Such individuals suffer from aggressive periodontitis with rapid periodontal breakdown [13].

### Kindler syndrome

The general findings include numerous genodermal disorders, blistering of skin following mild trauma, poikiloderma, thin wrinkled skin devoid of surface markings. Periodontal findings include tooth mobility, spontaneous gingival bleeding, earlier accelerated attachment loss and desquamative lesions of gingiva. Kindler syndrome has been proposed as a medically predisposing condition for destructive periodontal disease [14].

### Sjogren's syndrome

General findings include keratoconjunctivitis sicca, xerostomia and rheumatoid arthritis. There is a higher risk for periodontal breakdown of teeth in patients with Sjogren's syndrome [15].

### Marfan's syndrome

It is caused by impaired formation of fibrillin I. Patients often have a tall stature, aortic/mitral valve prolapse and ectopia lentis. The oral findings include severe periodontitis [16].

### Hyperimmunoglobulin E syndrome

It is a syndrome involving multiple systems including skeleton, connective tissue and immune system. Aggressive periodontitis and profuse gingival bleeding are oral clinical features [17].

### Cowden's syndrome

The general findings include papular lesions around nose, lips, ears, thyroid adenoma, breast cancer, and gastrointestinal polyps. It involves mutation in the PTEN gene on chromosome arm 10q. The periodontal findings include multiple papules on the gingiva giving an

appearance of a cobblestone pattern, gross dental caries and Periodontitis [18].

### Sturge-Weber syndrome

The general findings include brain calcifications, ocular disorders, hemiplegia, epilepsy and port wine stains confined to skin area supplied by the trigeminal nerve. The gingiva may show mild vascular hyperplasia or massive hemangiomas proliferation limited to one side. Gingival lesions may resemble pyogenic granuloma [19].

### Periodontal management

Patients with syndromes usually require psychological counselling for dental treatment. Complete explanation of the treatment options and procedure is essential. Written consent should be obtained from the parents and medical consent from the general physician. Chair positioning has to be proper as certain syndrome patients are prone to spinal cord injuries [5]. Initial cause related therapy should be completed first that includes patient education, motivation and oral hygiene instructions, complete supragingival and subgingival scaling and root planing, antimicrobial therapy if required and removal of any plaque retentive factors. If the candidate is suitable for surgical therapy, laser or electro cautery can be used for to minimize bleeding and post-operative complications. Post-operative antibiotics, analgesics and oral hygiene instructions are given. Some patients suffering from syndromes lack the manual dexterity to perform oral hygiene measures for themselves. In such cases oral hygiene instructions are given to family members or caretakers. Even after thorough periodontal treatment, periodontal destruction might continue in some patients. Implants can be considered in such cases however, due to lack of longitudinal studies in syndrome patients the survival rate of implants is a topic of debate.

### DISCUSSION

A case report of three cases of Sturge Weber syndrome having periodontal manifestations in the form of gingival enlargement has been reported. The author treated the cases with thorough scaling and root planing followed by gingivectomy by scalpel and laser [19]. A case report of two siblings suffering from Papillon Lefevre syndrome presented skin lesions and periodontal involvement with alveolar bone loss around multiple teeth. The patients were treated with non-surgical periodontal therapy and extraction of teeth with hopeless prognosis followed by maxillary and mandibular dentures [20]. There were similar case reports with the same treatment protocol [21,22]. A case report of a 12 year old girl with Chediak Higashi syndrome received initial periodontal treatment which resulted in an improvement in the clinical status and a stable periodontal status for a period of 9 years [13]. There are studies in the literature reporting an association between metabolic syndrome and periodontal disease [23,24]. A systematic review on periodontal disease in Down's syndrome patient shows

higher prevalence of periodontal disease in DS patients [25]. Case reports on Leukocyte adhesion deficiency syndrome-I also show early onset of periodontal disease which is not responsive to conventional non-surgical periodontal therapy [26].

### CONCLUSION

The association of periodontal disease with various syndromes questions the microbial etiology of dental plaque and presence of local factors to be essential for development of gingival inflammation and points towards a genetic basis of the disease. Poor oral hygiene with an altered structure of connective tissue in these syndromes can increase the severity of gingival and periodontal signs and symptoms. This occurrence of periodontitis in association with a syndrome is common in young adults that also suffer from dermal, ocular, psychological, neurological, skeletal and immune abnormalities. The contribution of syndromes to periodontal disease is minimal. However, knowledge and awareness of a periodontist regarding this association can lead to early diagnosis of such individuals leading to reduction in morbidity and an improvement in the general health and quality of life of patients.

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