Ehlers-Danlos syndrome is a genetically inherited connective tissue disorder where you

present with manifestations of hyperelastic skin, poor wound healing, joint hypermobility and tissue fragility. It is a disorder where your skin, muscles, and ligaments are not provided with sufficient tissue support.

Ehlers-Danlos syndrome was officially written about in 1657 by a surgeon called Van Meerkan although it was discovered by Hippocrates in 400 B.C.(1) It was not till the late 1930's where the disorder was accepted by the name of Ehlers-Danlos syndrome after Edward Ehlers and Henri-Alexandre Danlos, both whom were dermatologists.

Ehlers-Danlos syndrome is classified into six major types; classical, hypermobility, vascular, which tend to be the most common types we see and we also have kyphoscoliosis, arthrochalasia, and dermatosparaxis. (2) The 6 types are categorized in accordance to the signs and symptoms a person is manifesting with.

Ehlers-Danlos syndrome is genetically inherited; therefore one parent or both have the abnormal gene. There are two ways in which they can be inherited as. These are known as autosomal dominant and autosomal recessive. In autosomal dominant inheritance, one parent carries the abnormal gene passing it on to their offspring with a 50% chance of inheritance. In autosomal recessive, both parents are carriers of the abnormal gene and with a 25% chance their offspring will become carriers as well. (1)

The diagnosis of Ehlers-Danlos syndrome is evident with family history, medical examinations and definitive confirmation through biochemical, molecular and genetic testing. (3) With Ehlers-Danlos syndrome, other than the clinical diagnosis; there are physical characteristics that attribute to the syndrome.

Physical attributes of the syndrome include hyperelasticity of the skin; smooth, velvety, fragile; widened atrophic scars over pressure points such as elbows and knees; joint hypermobility; sprains, dislocations, subluxations; shaggy hair; narrow nasal bridge; foot deformities such as club feet; easy bruising; short stature and fingers. (3,5)

Intra-oral findings have also been observed such as absence of inferior labial and lingual frenula; (4) highly fragile mucosa; very soft tongue and almost 50% of individuals presenting with the syndrome can touch the tip of their nose with their tongue called Gorlins sign; the palate is deep and dome shaped. (5)

There is a significant connection between Ehlers-Danlos syndrome and oral health care providers. The dentist and dental hygienist must be very aware and cautious with providing dental treatment to patients with the syndrome. To avoid any complications during their dental visit, a thorough interview with the patient should be held along with taking their medical and family history. This is the safest and professional way to treat a patient.

There is a correlation between Ehlers-Danlos syndrome and periodontal disease also known as periodontitis Ehlers-Danlos syndrome type VIII. (2) When performing an oral assessment, there have been periodontal findings such as gingival bleeding, generalized membranous gingival enlargement and early-onset generalized periodontitis. (5,6) If there is no early intervention, it can result in bone loss, tooth mobility, and loss of primary and permanent teeth. (6)

In the oral cavity you may also see in patients with Ehlers-Danlos syndrome to have premolars and molars with deep fissures, long cusps, enamel hypoplasia and microdontia. Radiographically you may also observe pulp stones and root deformities. (5)

The recommended treatment plan first and foremost is to highly stress the importance of their oral hygiene. Using an electric toothbrush over a manual toothbrush can be recommended if patients are having restricted hand movement because of joint pain in the wrists. A fluoride mouth rinse should also be a homecare instruction for patients with Ehlers-Danlos syndrome because of their poor oral hygiene they are at high risk for caries, which also means they should have good nutritional habits to help decrease the possibility of caries to form.

To avoid any tissue trauma, such as excessive bleeding from fragile gums and mucosa, careful hand instrumentation would be preferable. Oral surgeons if possible should opt out from maxillofacial surgeries for Ehlers-Danlos syndrome patients due to chronic bleeding and poor wound healing. Orthodontic treatment should be done at a very gentle and soft paced rate taking into consideration that forceful movements damage further to the already fragile state the periodontal ligaments and mucosa are in. (5) Short duration of appointments would be optimal to prevent any trauma to TMJ and overall comfort for the patient.

Overall, it is our responsibility as professionals and oral health care providers to present our best judgment and treat patients accordingly to their medical conditions. This not only means we are doing our job but we are also making a difference in patients lives in providing them with the medical attention they deserve and very well need.

References

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