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Oral and Mandibular Manifestations in the Ehlers-Danlos Syndromes

FOR NON-EXPERTS

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The Ehlers-Danlos syndromes (EDS) are disorders that affect the connective tissues in the body. Several types of EDS have been identified. Mouth and jaw structures are affected differently in various types of EDS. These manifestations of EDS have been seen for many years, but newer diagnostic methods are shedding additional light on the challenges faced by EDS patients in the area of mouth and jaw disorders. Overly moveable jaw joint with associated consequences in EDS are noted. These features, their diagnosis, and treatment are presented.

Introduction

Mouth and jaw symptoms have been observed in all types of EDS patients. Collagen defects cause problems for mouth health including blood vessels, bone, teeth, gum tissue, nerve tissue, as well as the tendons and ligaments that retain facial/jaw

structures in position. These signs in EDS are often poorly recognized by doctors but are commonly reported by patients and impact pain, function, as well as quality of life. Here, we describe the mouth and jaw issues of EDS, diagnostic techniques, and treatment methods.

Appearance in the Soft Tissue of the Mouth

Structural collagen and its function is altered in all types of EDS. The lining inside the mouth is often thin and fragile. Easy wounding occurs with oral appliances. Poor wound healing and excessive bleeding is common with injury as well as dental procedures. Early onset gum disease is seen in a variety of EDS patients, especially the periodontal type (pEDS). Functional consequences of altered collagen could affect oxygen and nutrient delivery as well as likely affecting diffusion of nutrients and small molecules, which may have a role in the observed resistance to local anesthetics. The lack of frenula (the central connective tissue flaps between the gums and lips) has been noted in some studies. It has also been noted that up to 50% of EDS patients are able to touch their tongue to their nose (Gorlin sign) but it is unclear if the absence of the lingual frenulum has an impact on the presence of this sign.

Bone Structure and Teeth

In the EDS patient, tooth movement has been noted which may accelerate the gum recession. Dental therapy (orthodontics) is rapid due to accelerated tooth movement in EDS patients and is usually accomplished in one year or less. Unfortunately, rapid but mild relapses and tooth movement are noted usually by 18 months. Several tooth abnormalities have been noted among EDS patients. Rear teeth are reported to have high projections (cusps) and deep fissures. The roots may be abnormally shaped, joined and/or elongated, or the teeth may be poorly formed or absent.

The Temporomandibular Joint

The anatomy of the jaw joint (temporomandibular joint, TMJ) is complex. It should also be noted that the TMJ and its muscles and functions are intimately associated with functions of the head and neck. Therefore, the general anatomy of the head and neck must also be considered. An overly moveable TMJ has been linked to other overly moveable joints. Much like any joint in EDS, the TMJ often comes out of place. TMJ dislocation is noted to occur more often in women than in the general population which mirrors EDS. The TMJ can relocate once overextended but can cause pain and damage, limiting mobility. The muscles of chewing can be overused, contract

uncontrollably (spasm), and cause pain in other parts of the body, reducing function and quality of life.

Examination and Diagnosis of Temporomandibular Disorder (TMD)

The tracking of opening and chewing motions can be diagnostic of jaw joint disorder (temporomandibular disorder, TMD). Common symptoms of TMD include: (i) the jaw stuck to the side; (ii) limitation of opening; (iii) inability to chew; (iv) pain in front of the ear; (v) headaches in the temples or side of the jaw; (vi) tooth pain; (vii) inability to turn the head and/or neck; (viii) inability to get the back teeth together; (ix) fullness, itching, or pain in the ear(s); (x) “popping” or grating with movement of the TMJ. It is not uncommon that the patient suddenly dislocates their TMJ and thereafter has a limited mouth opening of 20–33 mm. This may be due to injury or stress, particularly the grinding of teeth and/or clenching of the jaws. When these habits are combined with EDS, the effects are amplified, particularly in those with an overly moveable neck.

The Relationship and Underlying Causes of TMD in EDS

Several studies have addressed overly moveable TMJ, other joints generally, and TMD with various conclusions. Research reported between 40% and 100% of patients with multiple types of headache and TMJ pain. A higher proportion of the EDS patients experienced hypermobile joints during extreme mouth opening, poor mouth opening ability when biting into thick food, clicking, grinding, and permanent locking of the jaw open and closed. Many of the EDS patients reported decreased mobility of the joint with age. Understanding the relationship between the head, neck, and jaw structures provides a key to pain management. The classic TMD headache is thought to be caused by muscle contractions triggered by things like stress and clenching the jaw.

Craniocervical Instability, Cervical Instability, Cervical Spine Disorders, and TMD

As early as 1934, researchers noted the relationship between an overly moveable head and neck (craniocervical instability, CCI), head and jaw problems, and headaches. A recent study examined the effect of TMD and positioning of the skull over the neck region. The authors found that 90% of the patients with cervical pain had TMD, concluding that the position of the spine (posture) impacted TMJ function. As the neck and upper spine are often involved in EDS patients, the interaction, recognition, and potential co-management should be considered.

Treatment and Management

Years of study of TMD, as well as advanced imaging techniques, have led to a deeper understanding of the syndrome, its causes, and treatment. Yet, while proper diagnosis and precise treatment of TMD is always complex, it is far more so in the EDS patient. Prevention of TMJ injury should be paramount. Posture, upper back, and neck issues need to be addressed. Lifestyle changes can include alteration of chewing patterns, diet, stress reduction techniques, and management of physical activities. Multiple treatments are available for management of pain and TMD associated problems, depending on the source and type of symptoms. Eliminating or minimizing muscle spasms is often the first step in reducing pain, and offers conservative treatment options which are appropriate for the EDS patient. Useful medications include muscle relaxants, mood elevators, anti-inflammatories, and pain medications. In the EDS patient, care must be taken to consider other medications and possible additional effects of any medication. *Botulinum* toxin to relax muscle or at trigger points can provide almost immediate relief for some patients. Physical therapy may be helpful; the therapist should understand the special needs of the EDS patient. Surgery should be limited to extreme cases.

Future Directions

Mouth and jaw problems are commonly encountered in the EDS patient but their true significance should be studied to establish the relationship between EDS and TMD. Additional study could also be informative for diagnosis and treatment, particularly in the area of head and face pain. Most importantly, the relationship of upper spine disorders to TMD in EDS patients requires further study. Recent studies have suggested a link between sleep-disordered breathing and EDS; this requires additional study. Additionally, further studies to document the occurrence of the oral frenula and Gorlin sign may be helpful.

Summary

Research has confirmed a variety of mouth and jaw signs associated with EDS. TMD and pain appear to be common in the EDS patient population. This relationship is logical given the nature of EDS and its effects on the multiple oral structures and collagen. The exact nature of this relationship merits further study, with the goal of providing more effective treatment.

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