2023 EDS ECHO SUMMIT

HYPERMOBILITY SPECTRUM DISORDERS

PRESENTATION

Dental Health and Orofacial Pain in HSD

SPEAKER

Rebecca Schaffer, DDS, DABOP

Assistant Professor, Advanced Care Clinic

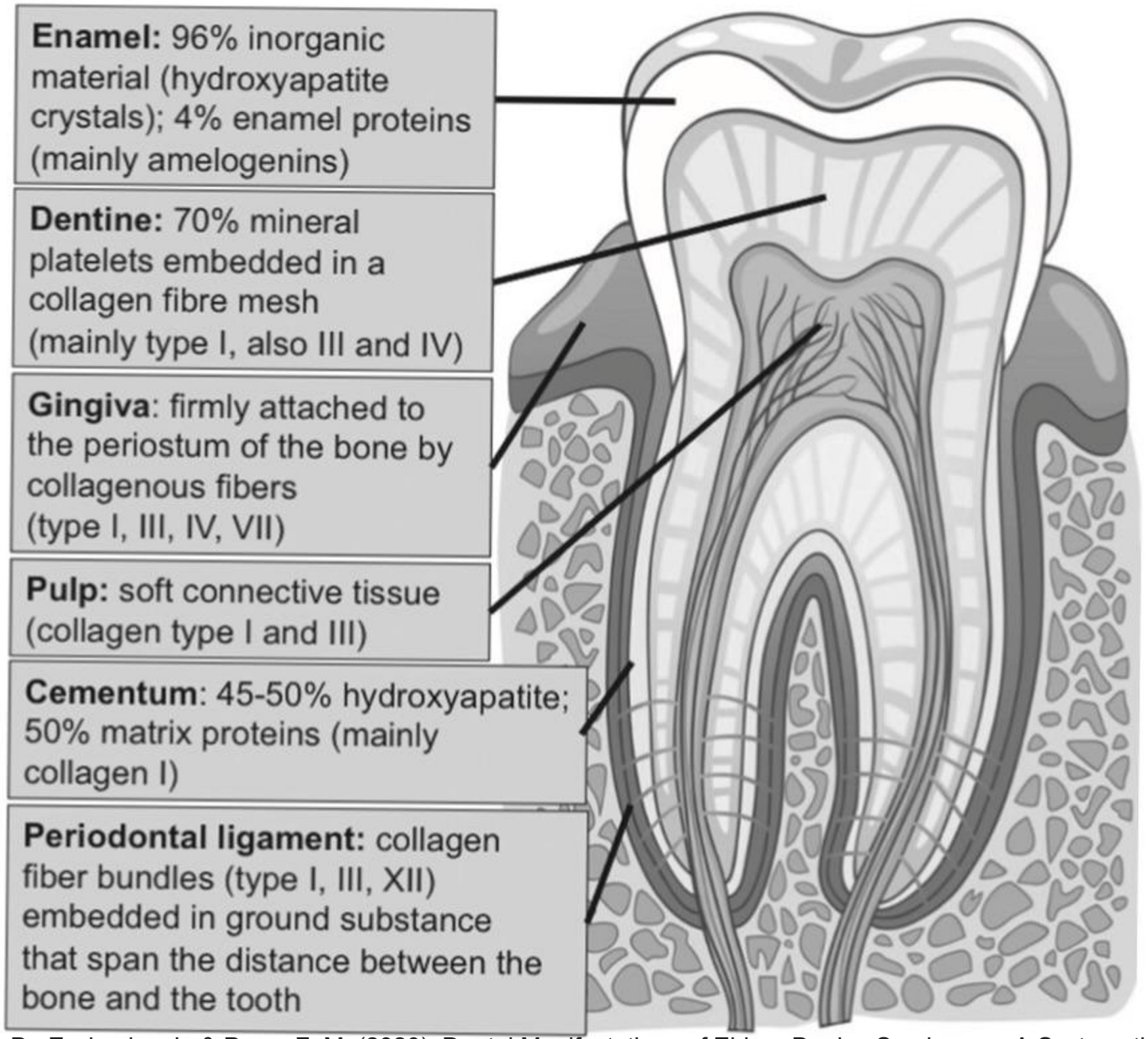
AT Still University, Arizona School of Dentistry & Oral Health

I have no disclosures

Learning Objectives

- Understand relationship between oral health and systemic health
- Improve communication among patients, oral health and other providers
- Empower patients to seek appropriate oral health care providers
- Explore the relationship between orofacial pain and HSD comorbidities

The tooth and its supporting tissues. The biology of dental tissues implies that tooth abnormalities might occur in various subtypes of Ehlers-Danlos syndrome.



Kapferer-Seebacher, I., Schnabl, D., Zschocke, J., & Pope, F. M. (2020). Dental Manifestations of Ehlers-Danlos Syndromes: A Systematic Review. *Acta dermato-venereologica*, *100*(7), adv00092. https://doi.org/10.2340/00015555-3428

Oral and Dental Manifestations of HSD/EDS

- Varies among different types of EDS
- Mucosal tissue is often thin and fragile
- Oral health compromised by chronic pain
- Pulp changes: stones, variable levels of calcification
- Dysplastic dentinal tubules

Oral and Dental Manifestations of HSD/EDS

- Poor response to local anesthesia
- Poor healing after oral surgery (extractions)
- Orthodontic treatment highly subject to relapse
- **Excessive translation of the mandible may not be effective to stop pharyngal collapse, which could be related to sleep-disordered breathing in EDS patients**

Local Anesthesia: Why doesn't it work?

- Online survey results
- 88% of people with EDS experienced inadequate pain prevention
- 33% of non EDS respondents experienced inadequate pain prevention
- Articaine(4%;1,100,000 epinephrine)was most successful in relieving pain
  - Still not reliably effective
- Lidocaine, which is the most commonly used anesthetic is least effective

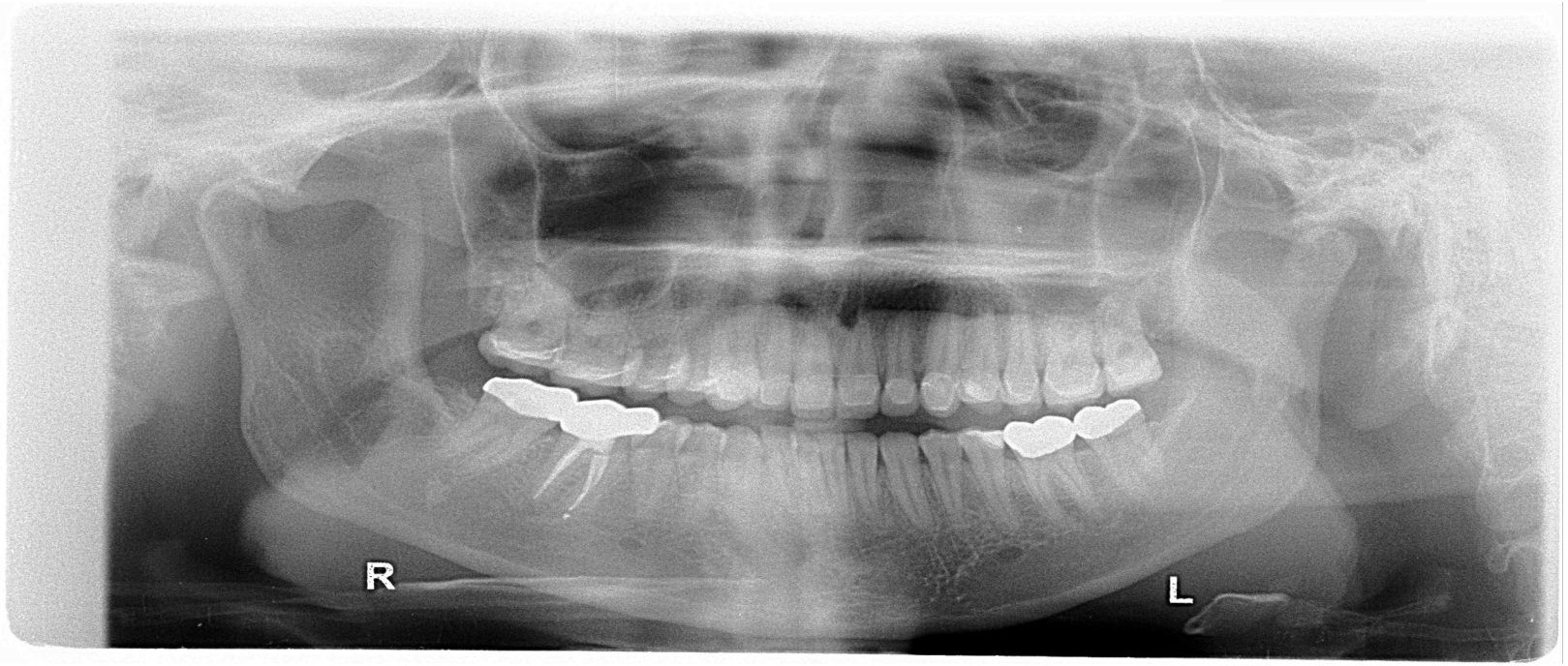
Schubart, J. R., Schaefer, E., Janicki, P., Adhikary, S. D., Schilling, A., Hakim, A. J., Bascom, R., Francomano, C. A., & Raj, S. R. (2019). Resistance to local anesthesia in people with the Ehlers-Danlos Syndromes presenting for dental surgery. *Journal of dental anesthesia and pain medicine*, *19*(5), 261–270. https://doi.org/10.17245/jdapm.2019.19.5.261

Local Anesthesia: Why doesn't it work?

- It is easier to achieve anesthesia on the maxilla (top Jaw)
- The mechanisms of local anesthetic resistance in HSD/EDS are not known
- Most dentists are unaware of the problem
- It can lead to fear and frustration

Case History: The Dentist's Role in Recognizing HSD/hEDS

- S:28 y/o female presents for comprehensive exam CC: ''check up and cleaning''
- Med hx: fainting spells, variable heart rate, numbness/tingling in extremities, dizziness, unusual headaches, chest pain when upset, hypothyroidism, acid reflux
- Pt reports she has been to multiple specialists who have diagnosed "anxiety" as a reason for fainting and other symptoms.
- Pt reports she has been falling and fainting all her life, and the dental crowns are a result of multiple broken teeth from various falls.
- Chairside Beighton score: 9/9

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LAS VEGAS
NEVADA

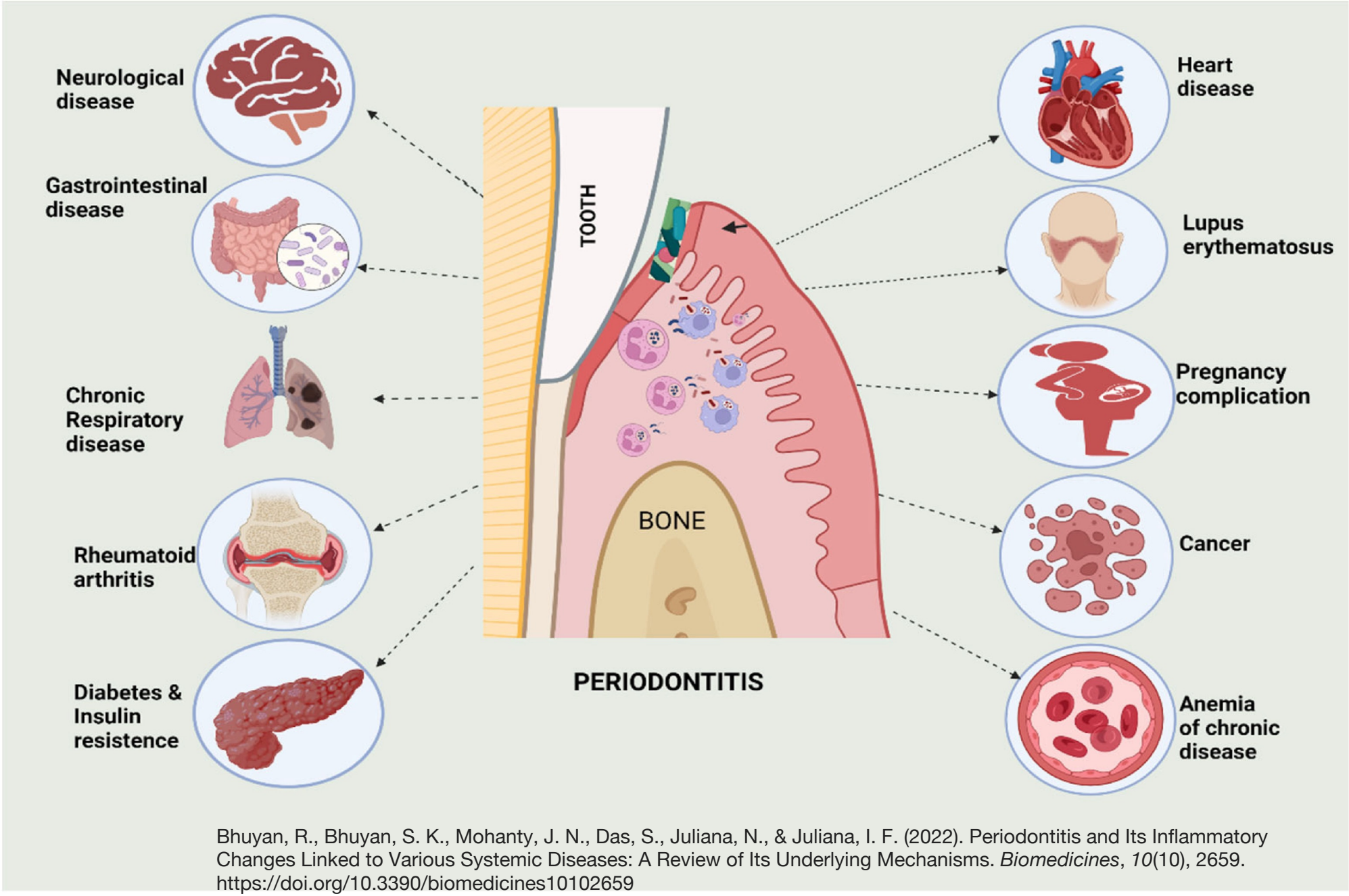
Periodontal Disease and Systemic Illness

- There is evidence of significantly higher levels of C-reactive protein (CRP) in periodontitis patients versus healthy controls
- Periodontitis patients exhibit significant endothelial dysfunction
- Positive association between periodontitis and cerebrovascular disease
- Joint disease (arthritis)

Sanz, M., Marco Del Castillo, A., Jepsen, S., Gonzalez-Juanatey, J. R., D'Aiuto, F., Bouchard, P., Chapple, I., Dietrich, T., Gotsman, I., Graziani, F., Herrera, D., Loos, B., Madianos, P., Michel, J. B., Perel, P., Pieske, B., Shapira, L., Shechter, M., Tonetti, M., Vlachopoulos, C., … Wimmer, G. (2020). Periodontitis and cardiovascular diseases: Consensus report. *Journal of clinical periodontology*, *47*(3), 268–288. https://doi.org/10.1111/jcpe.13189

The Mouth is the Gateway to the Body

- Neurological disease
- Gastrointestinal Disease
- Chronic respiratory disease
- Bidirectional cause and effect relationship with diabetes
- Autoimmune disease



Bhuyan, R., Bhuyan, S. K., Mohanty, J. N., Das, S., Juliana, N., & Juliana, I. F. (2022). Periodontitis and Its Inflammatory Changes Linked to Various Systemic Diseases: A Review of Its Underlying Mechanisms. *Biomedicines*, *10*(10), 2659. https://doi.org/10.3390/biomedicines10102659

Non-Odontogenic Pain

- After controlling for the effects of age, gender, and general joint diseases, hypermobile subjects (with four or more hypermobile joints on the 0–9 scale) had a higher risk for reproducible reciprocal clicking as an indicator for disk displacement with reduction (Odds Ratio (OR=1.68) compared with those subjects without hypermobile joints[1]

- Studies reported between 40% in one study and up to 100% in another, of patients with hSD/hEDS presenting with multiple types of headache and/or unilateral or bilateral TMJ pain

1.Hirsch, C., John, M. T., & Stang, A. (2008). Association between generalized joint hypermobility and signs and diagnoses of temporomandibular disorders. *European journal of oral sciences*, *116*(6), 525–530. https://doi.org/10.1111/j.1600-0722.2008.00581.x

Non-Odontogenic Pain

- The DC/TMD diagnoses myalgia, myofascial pain with referral, arthralgia, headache attributed to TMD, disc displacement disorders and degenerative joint disease occurred significantly more often in HSD/hEDS compared to controls (p = .000; p = .008; p = .003; p = .000; p = <.0001; p = .010, respectively) [2]

2. Bech, K., Fogh, F. M., Lauridsen, E. F., & Sonnesen, L. (2022). Temporomandibular disorders, bite force and osseous changes of the temporomandibular joints in patients with hypermobile Ehlers-Danlos syndrome compared to a healthy control group. *Journal of oral rehabilitation*, *49*(9), 872–883. https://doi.org/10.1111/joor.13348

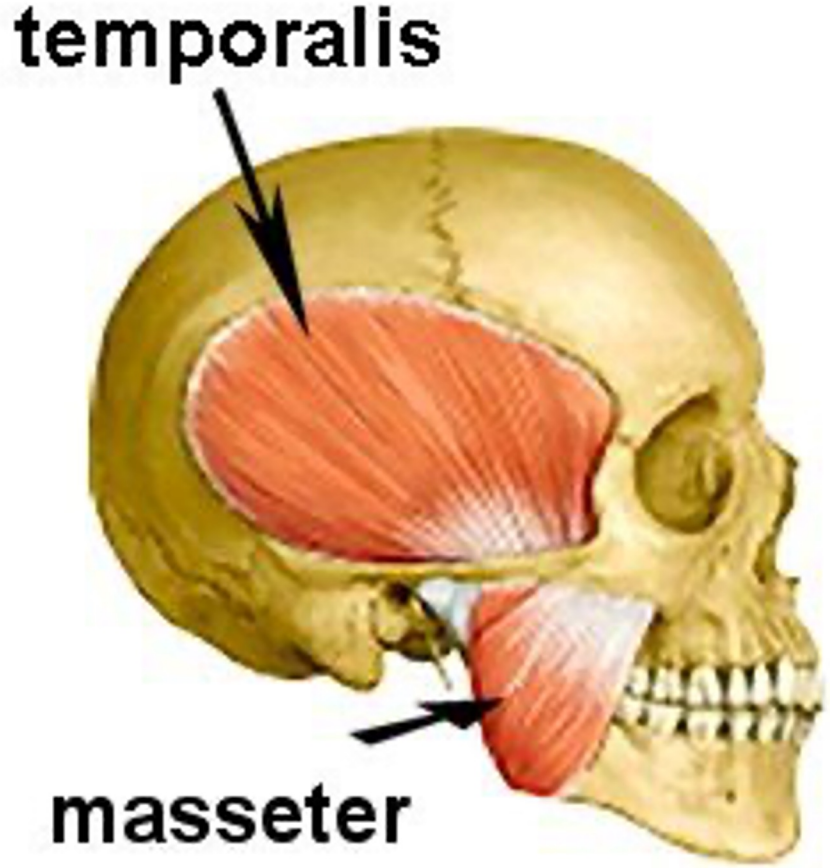
The muscles of mastication

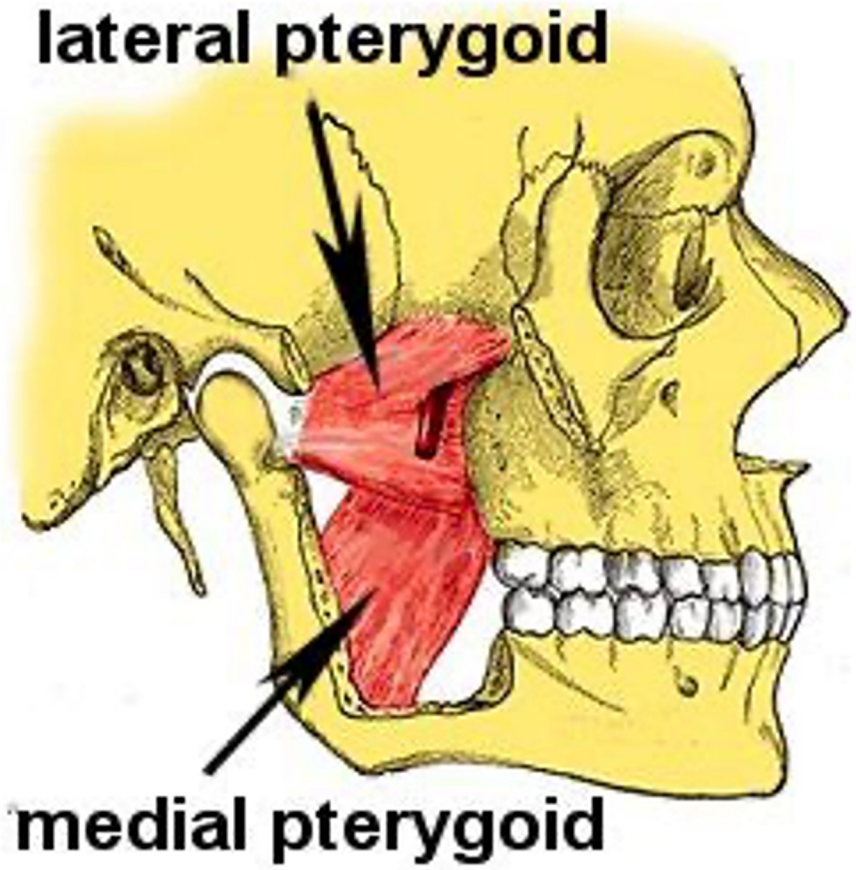
- Temporalis: elevates the mandible
- Masseter: elevates the mandible
- Medial pterygoid: elevates the mandible
- Lateral pterygoid: protrudes and depress the mandible

Accessory muscles

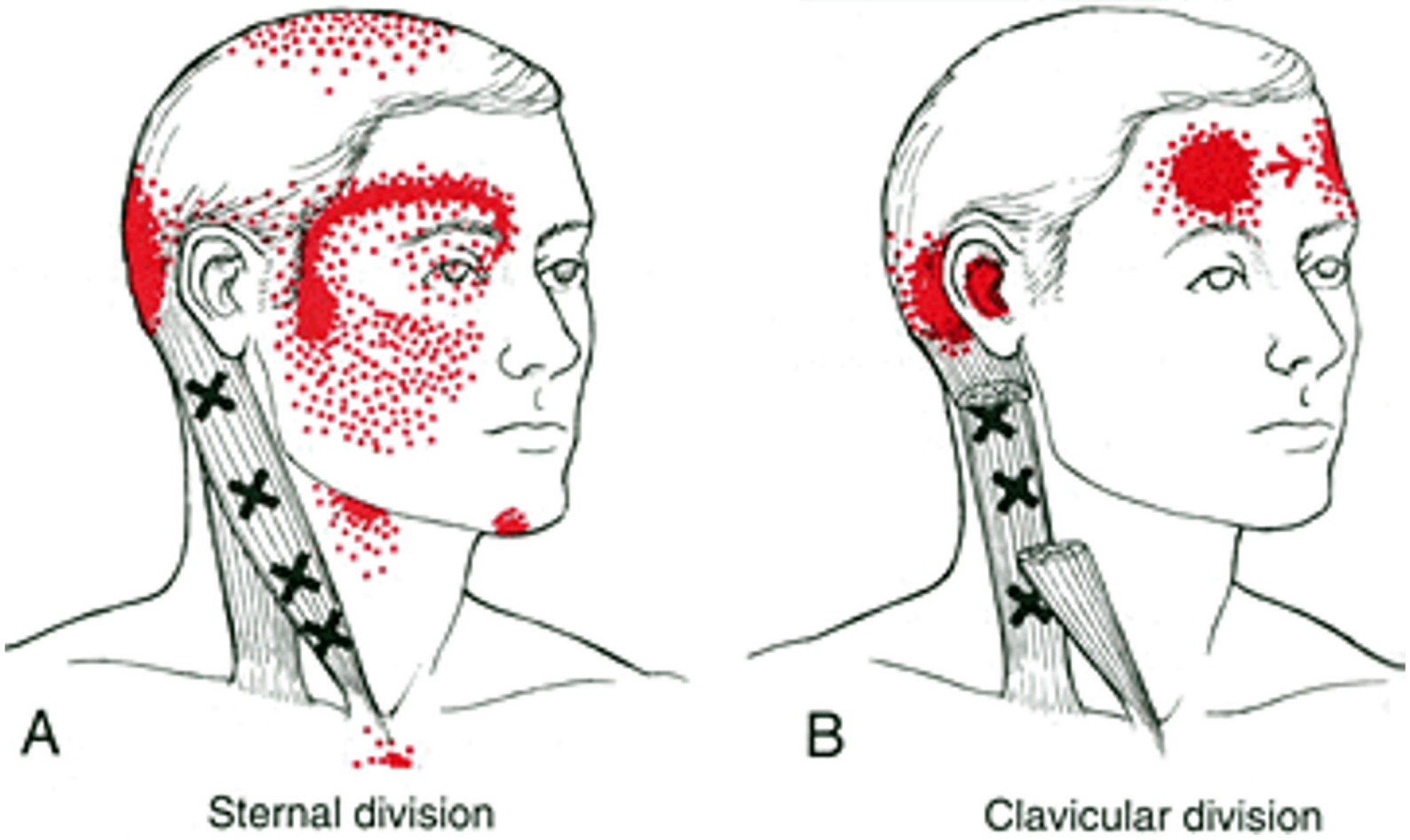
- Anterior/posterior digastric
- Buccinator
- Mylohyoid
- Geniohyoid
- Sternocleidomastoid
- Trapezius
- Posterior neck muscles

- Palpation of these muscles will delineate tender areas that generate myofascial pain referral patterns.


temporalis
masseter

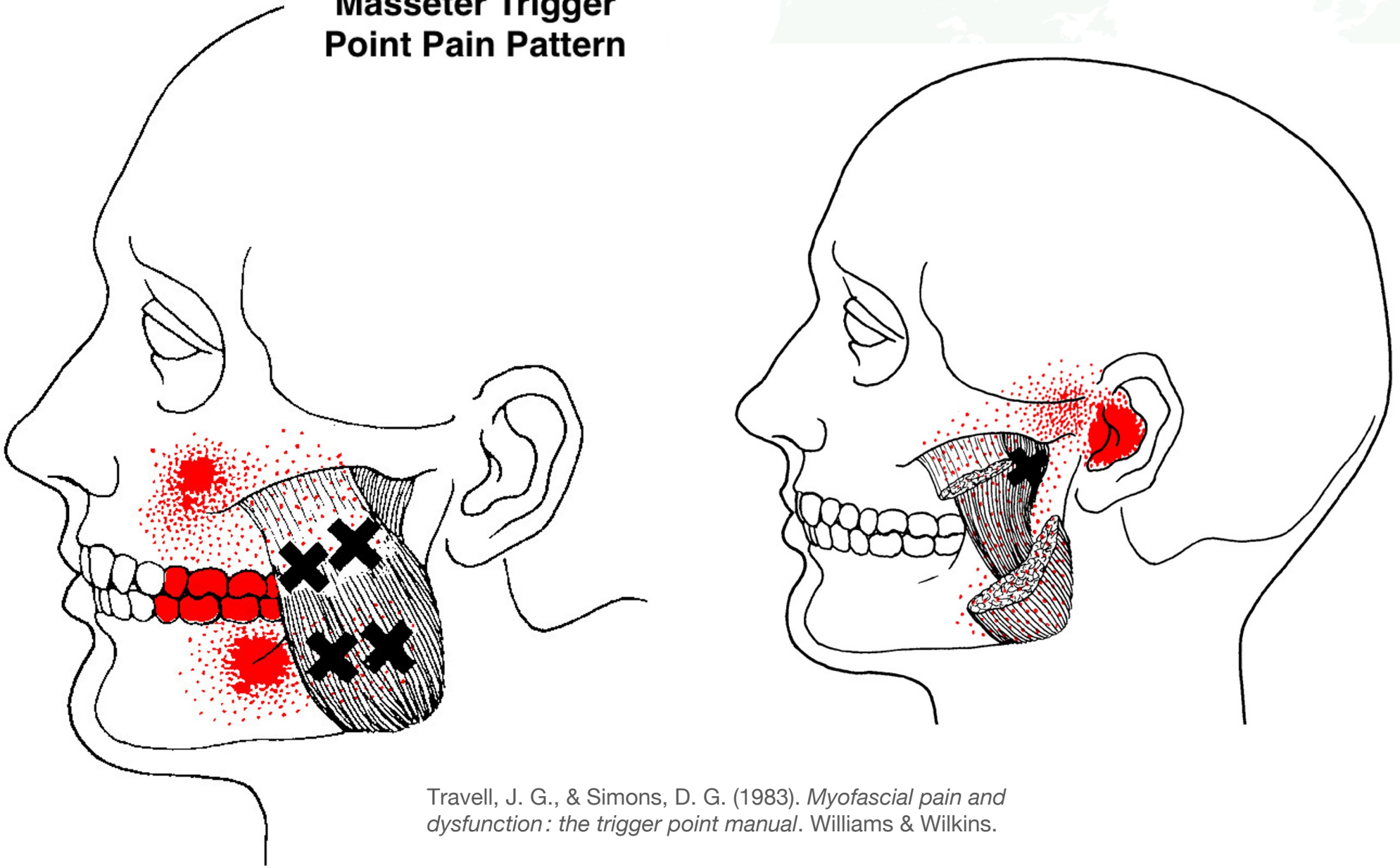

lateral pterygoid
medial pterygoid

Sternocleidomastoid



Travell, J. G., & Simons, D. G. (1983). *Myofascial pain and dysfunction: the trigger point manual*. Williams & Wilkins.

Masseter Trigger Point Pain Pattern



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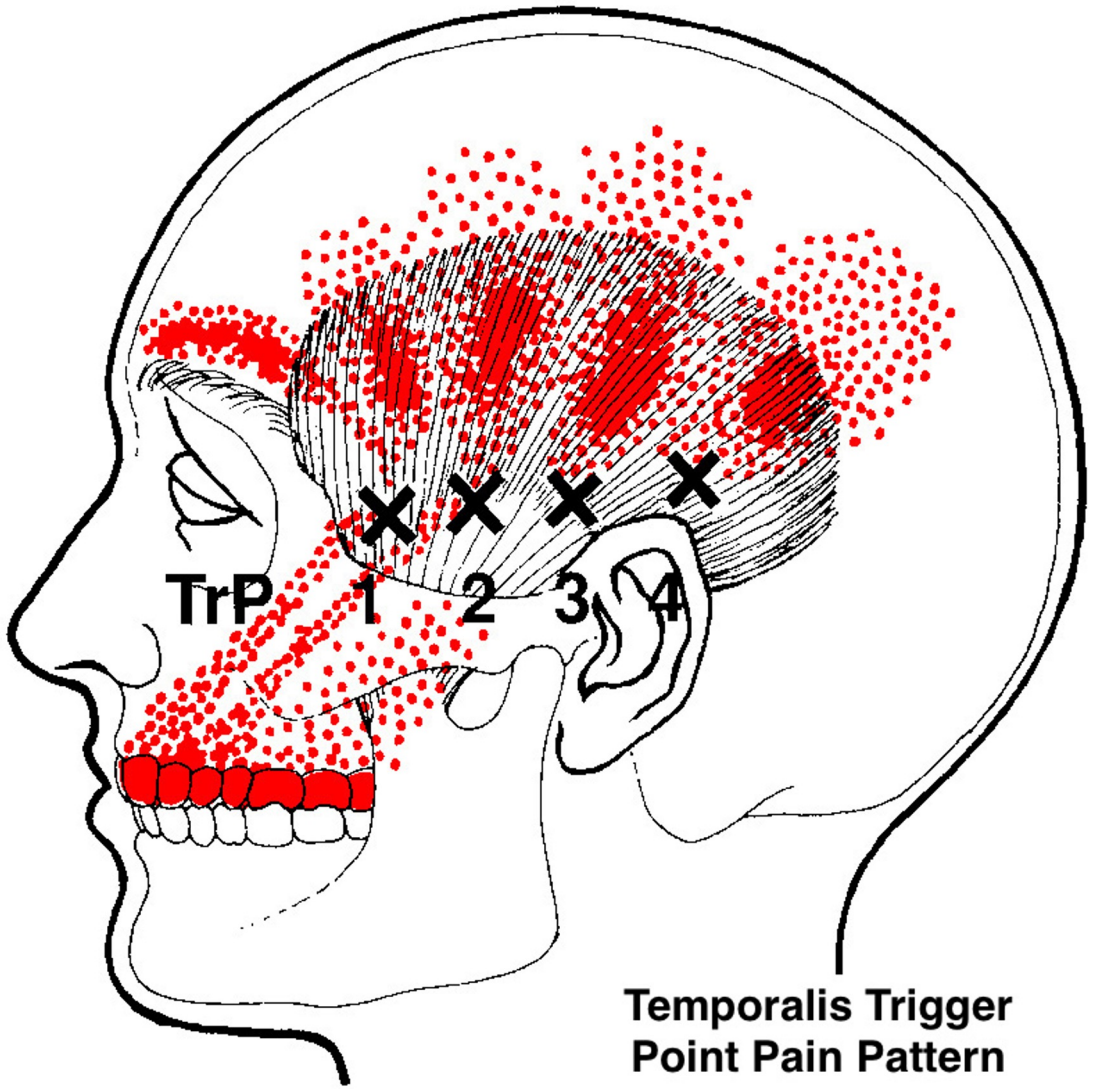

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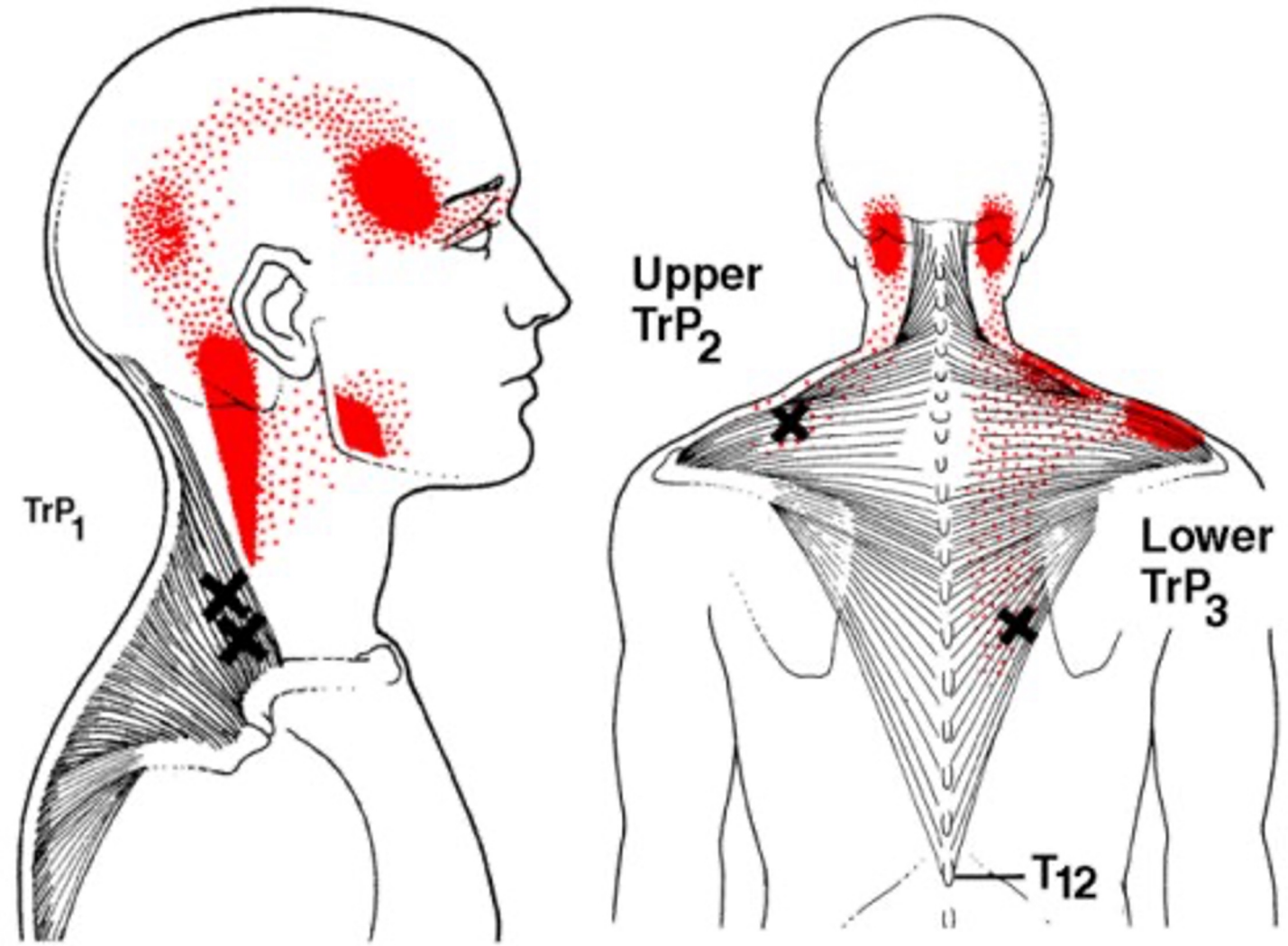

Travell, J. G., & Simons, D. G. (1983). *Myofascial pain and dysfunction: the trigger point manual*. Williams & Wilkins.

Trigeminal Nucleus Caudalis

- Longitudinally-shaped nucleus situated in the caudal pons and medulla
- Receives sensory afferents relating to pain and temperature from the orofacial region supplied by the trigeminal nerve
- Crosstalk with cervical nerves: source of referred pain
- By compressing the C2 nerve, **cervical spine instability can cause trigeminal neuralgia** and migraine headaches
- Stimulation of any of the upper cervical nerves (C1-C3) can also cause headache

An initial examination for TMD

- In the TMJs, the fibrocartilage structures, supporting ligaments, the disc and the retro discal tissues are composed mainly of collagen
- Joint noises
- Inflammation of TMJ structures
- Trigger points in muscle
- Referral patterns
- Wear patterns on teeth
- Postural disorders that impact TMD

John E. Mitakides (2018) The effect of Ehlers-Danlos syndromes on TMJ function and craniofacial pain, CRANIO®, 36:2, 71-72, DOI: 10.1080/08869634.2018.1435092

When to refer to OFP specialist

Do you have any of the following symptoms?

- Pain in the face, neck, jaw, temple, in front of the ear or in the ear in the last 1 month
- Any unresolved problems with headaches or migraines in the last 6 months
- Jaw problems that prevent or limit you from chewing or yawning or having your usual facial appearance
- Painful jaw click or pop when you open or close your mouth or when chewing

Exam:

- Palpation pain in the TMJ on opening / closing at ___ mm
- Severe pain on palpation to the masseters, temporalis,, sternocleidomastoids, upper trapezius
- Restricted mouth opening of less than 35mm (about 2 fingers width)

If none of the above exam items is positive, prediction is TMD negative. Otherwise, if even one exam items is positive, TMD is predicted and should be referred.

Prevention and Treatment of TMD

- Assuming TMJ hypermobility and generalized joint hypermobility increases the prevalence of TMD, it is reasonable to treat all EDS patients prophylactically.

- Prevention of TMJ injury should be paramount.

- Postural alignment, upper back ,cervical issues and jaw stabilization must be addressed as part of physical therapy treatment plans

- Lifestyle changes can include alteration of chewing patterns, diet, stress reduction techniques, and management of physical activities.
  - Heat/ice application
  - Diaphragmatic breathing
  - Mindful relaxation techniques
  - Jaw stabilization devices?

Looking Ahead

- Develop evidence based protocols for
  - Routine dental visits
  - Nutritional guidance
  - Oral hygiene modifications
  - Physical therapy for cervical/orofacial pain
- Improve interprofessional communication
- MCAS and periodontal disease
- Cause(s) of resistance to local anesthesia
- Empower patients to speak up about their issues and develop effective communication tools

Empowering our Patients: Checklist for Dental Visits

- I have _____hypermobility/Ehlers Danlos Syndrome
- HSD/EDS is a condition which affects collagen and connective tissue
- I do not respond to lidocaine. It is a symptom of my condition.
  - Please use 4% articaine to get me numb
  - I may need more than one injection
- I often have pain for several days even after routine procedures_____
- I have difficulty healing after extractions_____
- Please support my neck with a pillow___
- Please don't put the chair back all the way____
- I get dizzy or faint if I stand up too fast___
- My gums bleed a lot___
- I bleed/bruise easily____
- My jaw pops/clicks/locks open____
- I have a lot of jaw pain
- I get frequent headaches___
- Please communicate with my doctor before any surgical or difficult procedures_____
- I cannot tolerate long appointments____
  - It hurts my jaw___
  - It hurts my neck___

TMD Exam - Brief Overview

https://youtu.be/z1yarO0rLZk

Conclusion: Connecting the Dots

- Oral health = overall health
- We are all responsible for encouraging oral health in our patients
- HSD/EDS = higher risk for oral/TMJ diseases and their consequences
- Better understanding & attention to quality of life improved outcomes
- Limited access to appropriate care

MORE HSD/EDS Awareness, Advocacy & Research is Needed

Thank you for listening

rschaffer@atsu.edu