



Hyaluronidase Injections for the Treatment of Microstomia Secondary to Systemic Sclerosis



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Background

- Systemic Sclerosis (SSc) is a progressive autoimmune disease characterized by cutaneous fibrosis
- In Limited cutaneous systemic sclerosis (lcSSc, formerly known as CREST syndrome), cutaneous involvement can be severe on the distal extremities, face, and neck
- Limited scleroderma commonly involves the perioral skin, leading to a narrowing of the oral aperture termed **microstomia**

Our Patient

- Our patient, Ms. M, presented to clinic with a longstanding history of limited scleroderma with myositis overlap syndrome
- Upon presentation, she was already suffering from sclerodactyly, calcinosis cutis, Raynauds, and microstomia
- Despite systemic treatment (Mycophenolate, IVIG) her disease continued to greatly affect her life



“What bothers you the most?”

Treating Microstomia in Limited SSc

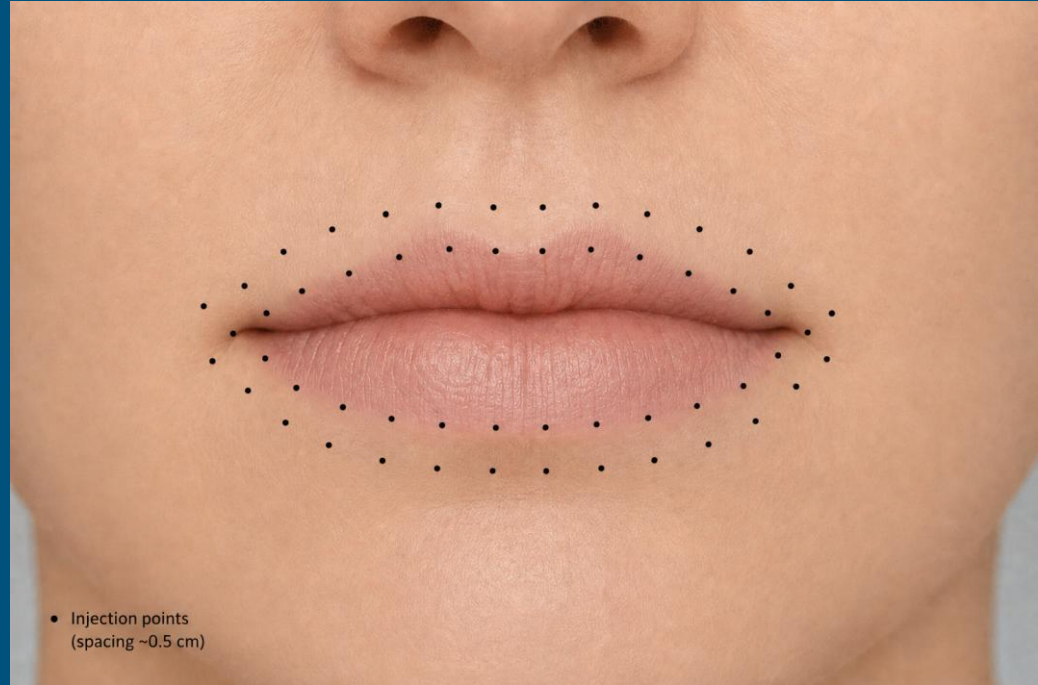
- Despite the impact microstomia has on patients' quality of life, there are no FDA approved treatments for longstanding scleroderma-related microstomia

Treating Microstomia in Limited SSc

- Limited case series and reports have demonstrated the efficacy of local, perioral subcutaneous injections of hyaluronidase to treat scleroderma-associated microstomia [1-4]
- Proposed mechanism: exogenous hyaluronidase degrades hyaluronic acid, reducing collagen deposition and promoting lysis of fibrinous material [1-2]

Technique

1. Nerve blocks performed of the infraorbital and mental nerves bilaterally
2. 1.2 cc vial (150 units) of hyaluronidase is diluted with 4.8 cc of normal saline
3. 6 cc total are drawn up into syringes with 30G needles
4. 1.5 cc injected into each quadrant of the upper and lower lip



Risks/Adverse Effects

- The number one side effect we saw was bruising
 - Ms. M happily showed us her “handlebar mustache” from her first round of injections
- Other side effects to counsel patients on are swelling, bleeding, and injection pain
- Overall, very safe and well tolerated

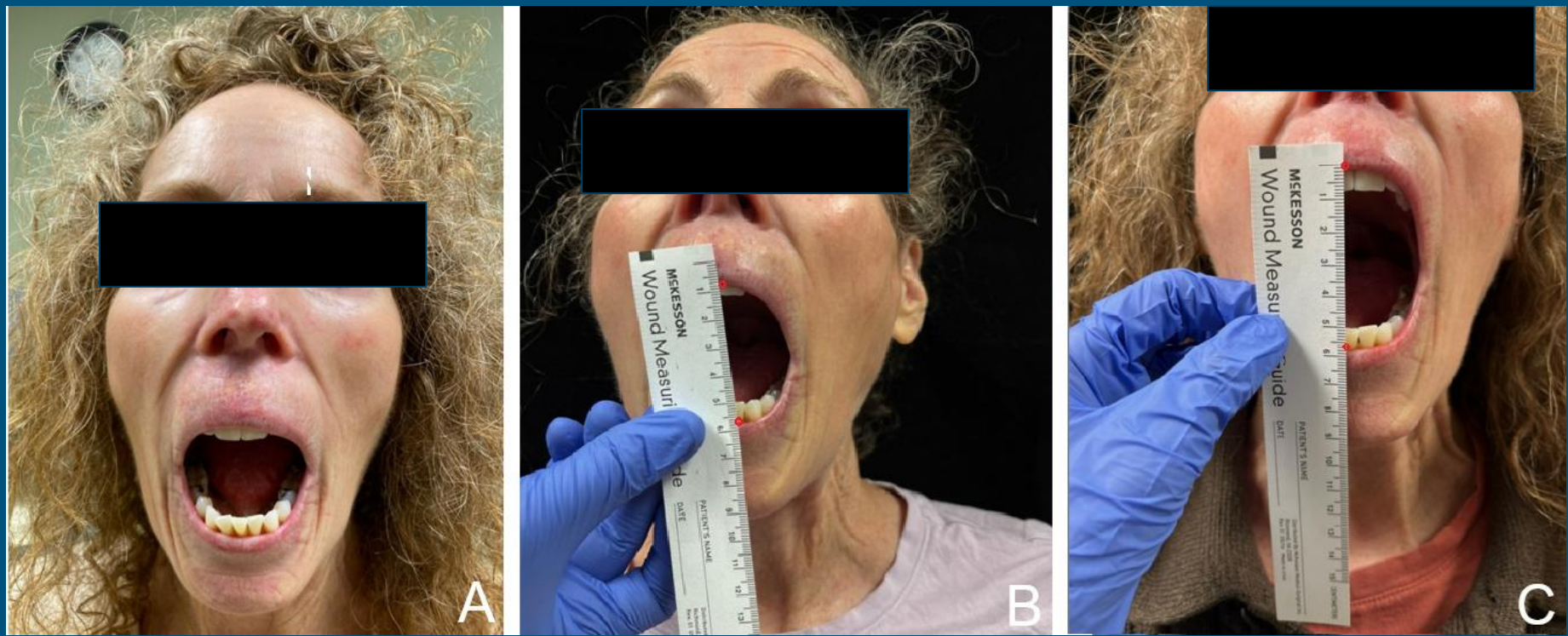


Figure 1: Baseline (A), after one round of injections (B) and after four rounds of injections (C)

Treatment	Vertical oral aperture (cm)	Horizontal oral aperture (cm)
Treatment 1	4.8	4.4
Treatment 2*	4.5	4.4
Treatment 3	5.3	4.4
Treatment 4	5.4	4.5
Treatment 5	5.7	4.6
Treatment 6	5.9	4.8
Treatment 7	5.8	5.0
Treatment 8	5.7	4.8
Treatment 9	5.7	5.1

Total Improvement:
Vertical: 1.1 cm
Horizontal: 0.7 cm

*Patient had dental procedure that limited oral aperture opening immediately prior to visit

Why do we keep going?

“What bothers you the most?”

“I have my smile back”



* This is the one case of increased wrinkles and a gummy smile I will brag about as a dermatologist.

Take Home Points

- 1) The most exponential objective improvement was seen within the first 4-6 visits
- 2) Even after measurements objectively stagnate, patients may experience subjective improvement due to “softening” of the skin surrounding the oral aperture
- 3) Treatment is simple, relatively risk-free, and well tolerated with nerve blocks
- 4) **Remember to ask your patients what bothers them - it's often different than what bothers you**

Questions?

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References

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