

MYASTHENIA GRAVIS AND DYSPHAGIA

Sheila Harkaway, MS, CCC-SLP
Michigan Medicine

DISCLOSURE

- My name is Sheila Harkaway. Today I will be talking to you about dysphagia management in patients with Myasthenia Gravis. I work at Michigan Medicine. I receive a salary from Michigan Medicine. I have no other relevant financial disclosures.

LEARNING OBJECTIVES

- Understanding what Myasthenia Gravis is
- Increasing familiarity with some of the medical interventions used in patients with Myasthenia Gravis
- Understanding the importance of a comprehensive motor speech evaluation with this population
- Understanding the Speech-Language Pathologists role in evaluation and treatment of patient with Myasthenia Gravis

WHAT IS MYASTHENIA GRAVIS?

- An autoimmune neuromuscular disease leading to fluctuating muscle weakness and fatigability
- Caused by a disruption in the normal communication between nerves and muscles (neuromuscular junction)
 - The proteins include AChR and less commonly muscle-specific tyrosine kinase (MuSK) involved in AChR clustering.
- Characterized by weakness and rapid fatigue of skeletal muscles
- Bulbar muscle involvement and dysphagia are commonly seen in patients with Myasthenia Gravis.

HOW IS MYASTHENIA GRAVIS DIAGNOSED?

- Ice Pack Test
- Blood Analysis
- Repetitive Nerve Stimulation
- Single-Fiber Electromyography (EMG)

COMMON MEDICAL INTERVENTION

- Pyridostigmine
 - Mestinon
 - Orally active cholinesterase inhibitor
- Thymectomy
 - Surgical removal of the thymus gland has been found to decrease symptoms in as many as 70% of patients who have thymomas or dysplasia of the thymus gland
- Rituximab
 - An antibody designed to interact with a specific target against the CD-20 that is present in B-cells.

COMMON MEDICAL INTERVENTION

- Intravenous immunoglobulin G infusions (IVIg)
 - IVIG is a blood product composed of purified and concentrated immunoglobulins (antibodies) from the pooled plasma of many healthy donors.
 - It binds circulating antibodies, thereby promoting Ach availability and muscle function.
 - Given 2g/kg over 5 consecutive days
- Plasmapheresis (aka therapeutic plasma exchange, TPE)
 - Plasmapheresis separates the plasma from other components in the blood, especially the red blood cells. The patient's plasma is exchanged with donor plasma or albumin solution.
 - The goal of plasmapheresis is the removal of receptor antibodies from the circulation.
 - 3-5 treatments administered every other day

PATIENT WITH REFRACTORY MYASTHENIA GRAVIS

- Solaris
 - Designed to block the terminal complement cascade
 - Only used on patients who have tried all other treatment options with no improvement

CLINICAL EVALUATION OF PATIENT WITH MYASTHENIA GRAVIS

- Oral motor evaluation
- Motor speech evaluation
- Clinical swallow evaluation

ORAL MOTOR EVALUATION

- Facial musculature (ptosis)
- Fatigability with repetitive movement
- Palatal elevation
- Gag response
- Secretion management

MOTOR SPEECH EVALUATION

Areas of Focus:

- Respiratory support
- Phonation/voice
- Articulation
- Resonance
- Prosody
- Intelligibility

Tasks Performed

- Sustained /a/
- Oral diadokokinetics
- Stress Testing
- Conversational Speech
- Nasal Emission Mirror

CLINICAL SWALLOW EVALUATION

- Secretion management
- Mastication
- Oral residue
- Excess swallows
- Fatigability with continuous trials
- Overt clinical signs/symptoms of penetration/aspiration

WHY FORMAL IMAGING?

- A study by Kluin et. al, looked at dysphagia in elderly men with Myasthenia Gravis.
 - All patients had pharyngeal phase greater than oral phase dysfunction.
 - All patients had decreased pharyngeal mobility demonstrated by residue in the valleculae and pyriform sinuses bilaterally
 - 7 patients had episodes of penetration due to overflow from residue
 - In 5 patients there was subsequent episodes of aspiration due to the overflow with no reflexive cough
 - There was an impression of the cricopharyngeus muscle in five patients.

CRICOPHARYNGEAL DYSFUNCTION

- A case study by R. Rison, 2009, looked at reversible oropharyngeal dysphagia secondary to cricopharyngeal sphincter achalasia in a patient with myasthenia gravis.
 - 83 year old man with a nine year history of MG and dysphagia
 - Last major MG crises was 8 years prior (responded to plasmapheresis)
 - On exam he had a mild dysarthria. No ptosis, facial symmetry or fatigability.
 - Initiated IVIG and plasmapheresis, no improvement noted
 - VFSS was completed and revealed a prominent cricopharyngeus sphincter with incomplete relaxation
 - Underwent cricopharyngeal dilation and dysphagia significantly improved.

EXERCISE AND MYASTHENIA GRAVIS

- Myasthenia Gravis is caused by a disruption in the normal communication between nerves and muscle leading to muscle fatigue.
- Electrical stimulation is **NOT** appropriate for patients with Myasthenia Gravis.
 - Electrical stimulation involves the application of current to stimulate nerves or nerve endings that innervate muscles or are sensory.

INTERVENTION FOR DYSPHAGIA

- Dietary Modifications
- Environmental Changes
- Alternative means of nutrition/hydration/medication.

CASE STUDIES

- Case Study #1
- 66 year old woman with AChR antibody positive Myasthenia Gravis
- Admitted with dysphagia. Reported that she had not been able to eat for a few days prior to admission.

CASE STUDIES

- **Oral Motor Examination:**
Facial/labial: Bilateral ptosis. Reduced labial ROM for pucker/smile. Reduced strength to resistance.
Mandibular: Reduced ROM for opening/lateralization. Reduced strength to resistance.
Lingual: Protrusion midline. Reduced ROM and strength to resistance.
Palatal: Reduced elevation bilaterally. Diminished gag.
-
- **Motor Speech Perceptual Examination:**
Respiratory support for speech: Sustained phonation of /a/: 18 seconds (norm is 16-20).
Phonation/Voice: Hypophonic and breathy.
Articulation: Moderate imprecision. Fatigable.
Resonance Severe hypernasality.
Prosody: Short utterances.
Speech intelligibility was judged to be 70%.

CASE STUDIES

- **Oral Agility:** Oral diadochokinetic rates were attempted for 5-second trials. Disturbances noted in voice, resonance, articulation and rate.

Test	Patient performance	Norm
/pa/	14	28
/ta/	12	28
/ka/	9	25
/pataka/	4	8-10

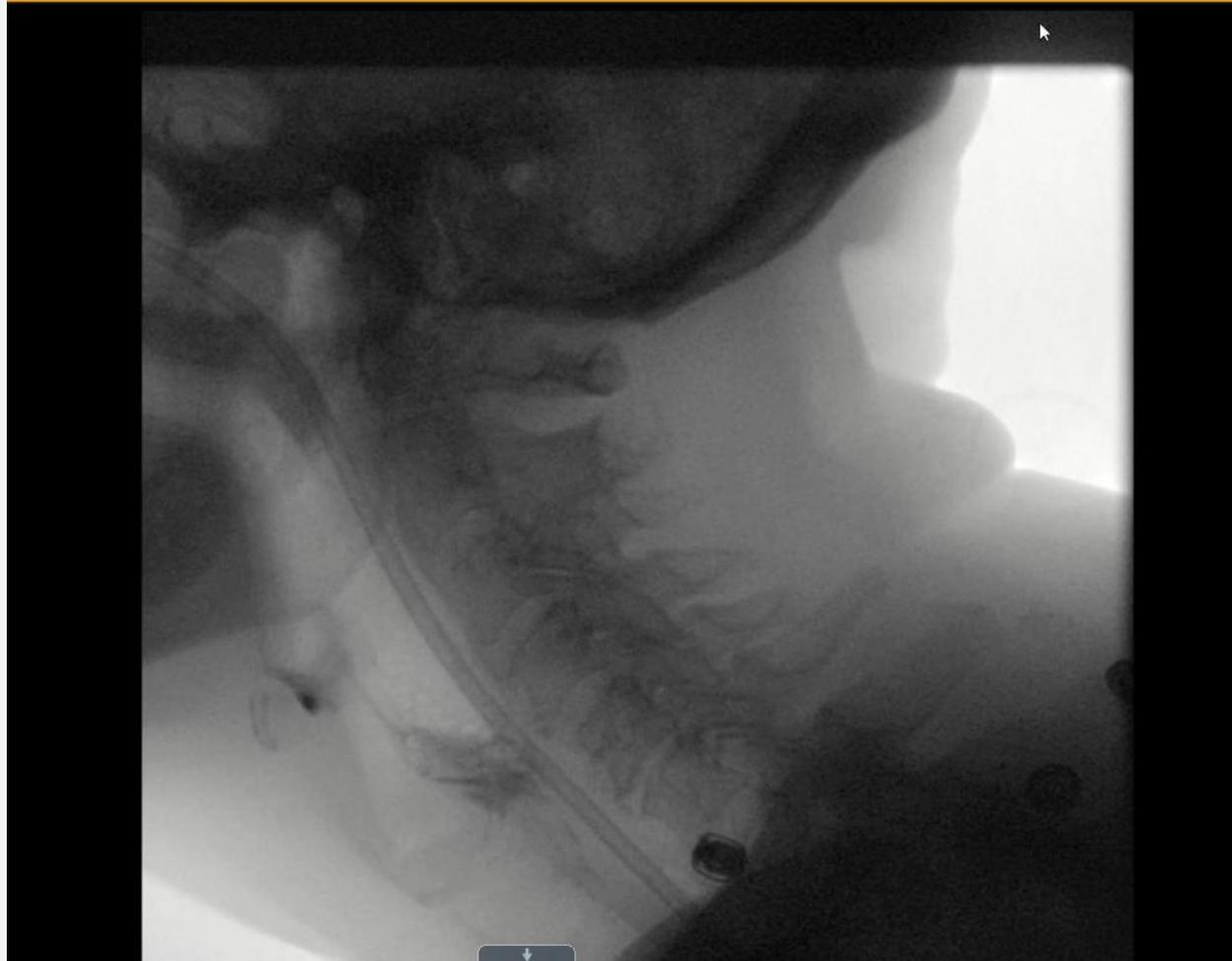
CASE STUDIES

- **Swallowing:**
Trials included:
Ice chips
Thin liquid (water) via spoon
- Performance notable for:
Reduced labial seal; no overt anterior spill.
Reduced ROM for mastication.
Swallow onset appeared timely.
Multiple swallows per bolus.
Immediate wet vocal quality and weak throat clear.

VFSS-THIN LIQUID VIA SPOON



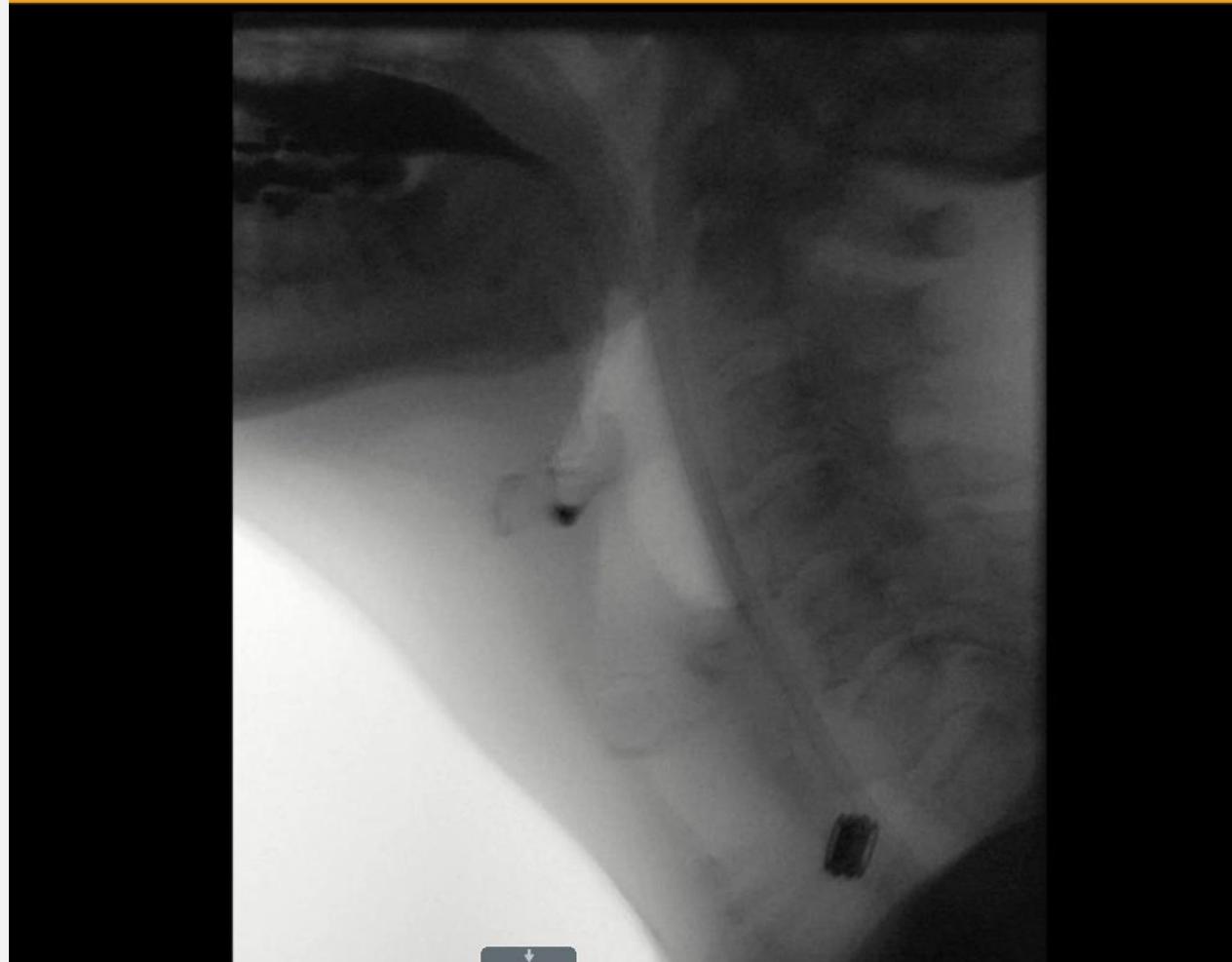
VFSS-THIN LIQUID VIA CUP



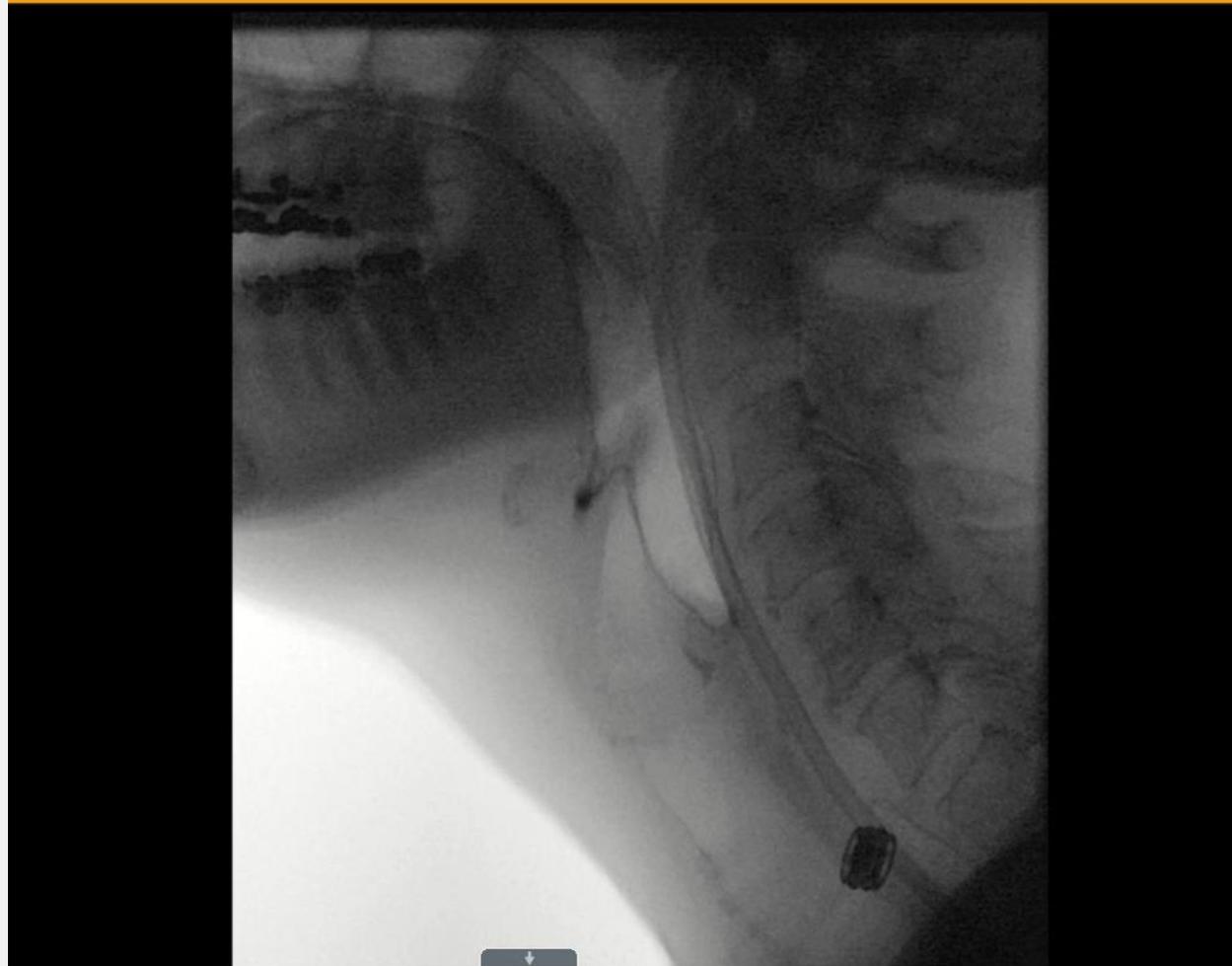
VFSS-THICK LIQUID VIA SPOON



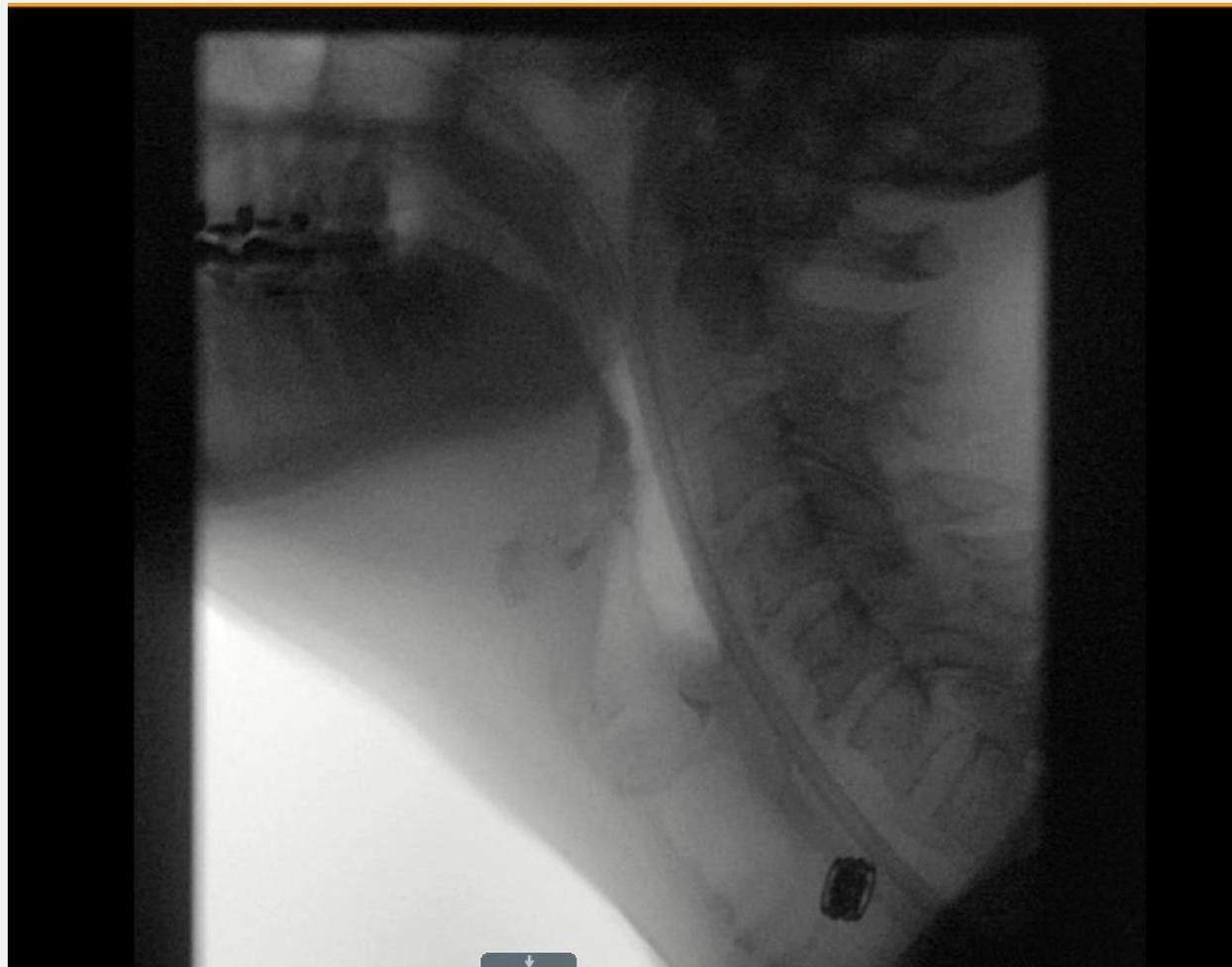
VFSS FOLLOWING PLASMAPHERESIS THIN LIQUID



VFSS FOLLOWING PLASMAPHERESIS PUREE



VFSS FOLLOWING PLASMAPHERESIS SOLID



CASE STUDIES

- Case Study #2
- 68-year-old man admitted to acute rehabilitation following CHF exacerbation
- PMH
 - Chronic diastolic HF, moderate MR, bicuspid aortic valve with moderate aortic regurgitation, CAD and remote history of TBI in 1970's
- On a regular diet

CASE STUDIES

- On my initial evaluation, here were my oral motor and motor speech findings:

Oral Motor Examination:

Facial/labial: Symmetrical, adequate ROM/strength to resistance.

Mandibular: Adequate strength to resistance and ROM for opening/lateralization.

Lingual: Midline protrusion. Adequate strength to resistance/ROM.

Palatal: Symmetrical. Fair elevation. Bilateral gag response.

Cough: Not elicited.

- **Motor Speech Perceptual Examination:**

Phonation/Voice: Slightly harsh.

Articulation: Mild imprecision.

Resonance: Unremarkable.

Prosody: Slow rate.

Speech intelligibility was judged to be 100%.

CASE STUDIES

- 6 days later I received a referral to evaluate swallowing due to patient noticing increased difficulty. Here are my oral motor and motor speech findings from that examination:
- **Oral Motor Examination:**
Facial/labial: Trace left labial droop at rest and with activation. Reduced labial strength bilaterally.
Mandibular: Fair ROM for opening/lateralization. Fair strength to resistance.
Lingual: Slight deviation to the left with protrusion. Reduced labial ROM. Bilateral labial weakness.
Palatal: Reduced elevation bilaterally. Diminished gag.
- **Motor Speech Perceptual Examination:**
Respiratory support for speech: Sustained phonation of /a/: 12 seconds (norm is 16-20).
Phonation/Voice: Harsh, hoarse and hypophonic.
Articulation: Severe imprecision.
Resonance: Continuous hypernasality.
Prosody: Short utterances.
Speech intelligibility was judged to be 60%.

CASE STUDIES

- **Swallowing:**
Trials included:
Ice chips
Thin liquid (water) via spoon, cup, and straw
Puree (applesauce)
Further trials deferred due to concern for aspiration.
- Performance notable for:
Fair labial seal; no noted anterior spill.
Swallow onset appeared timely.
Multiple swallows per bolus.
Immediate coughing noted with all trials.

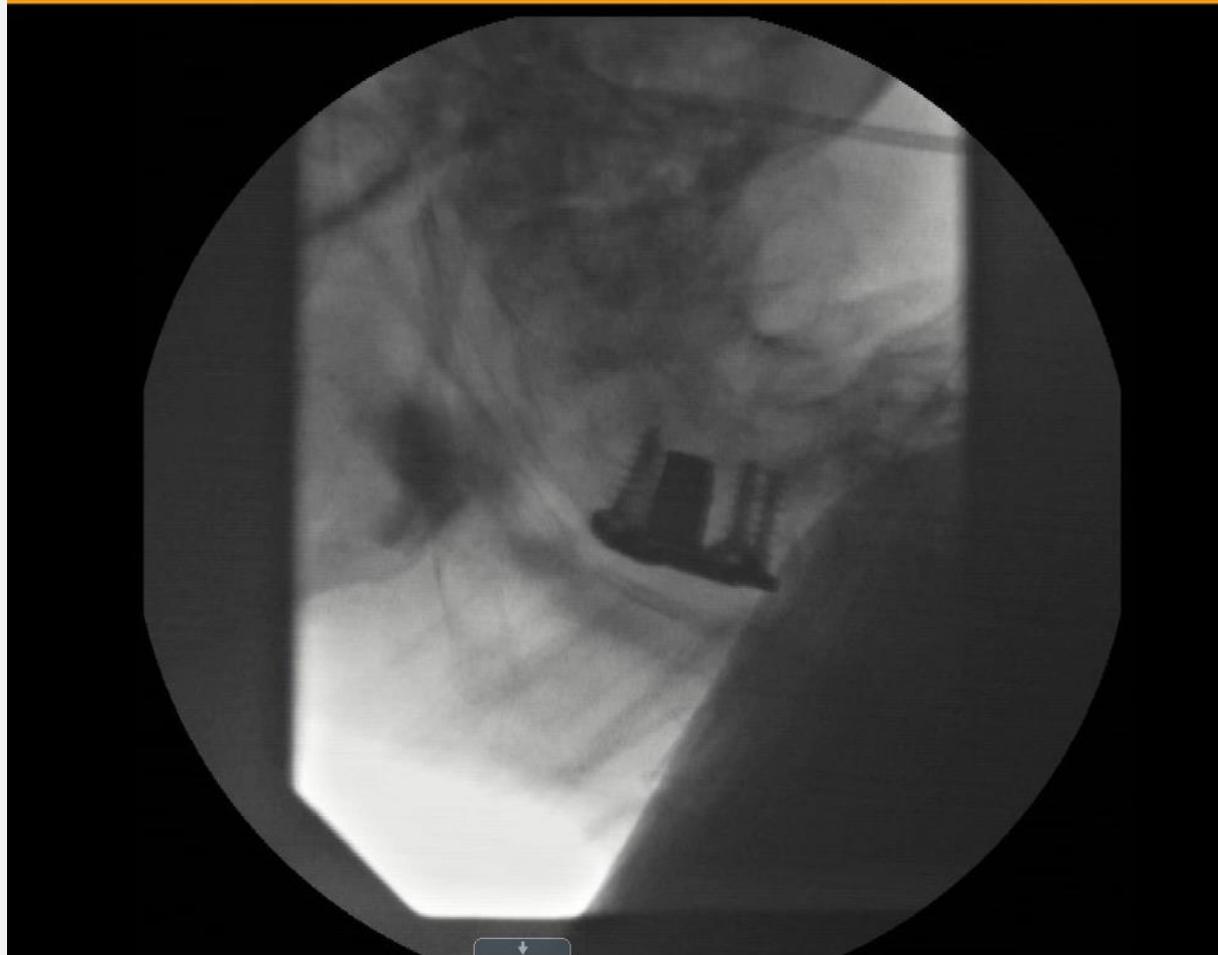
VFSS-THIN FROM A SPOON



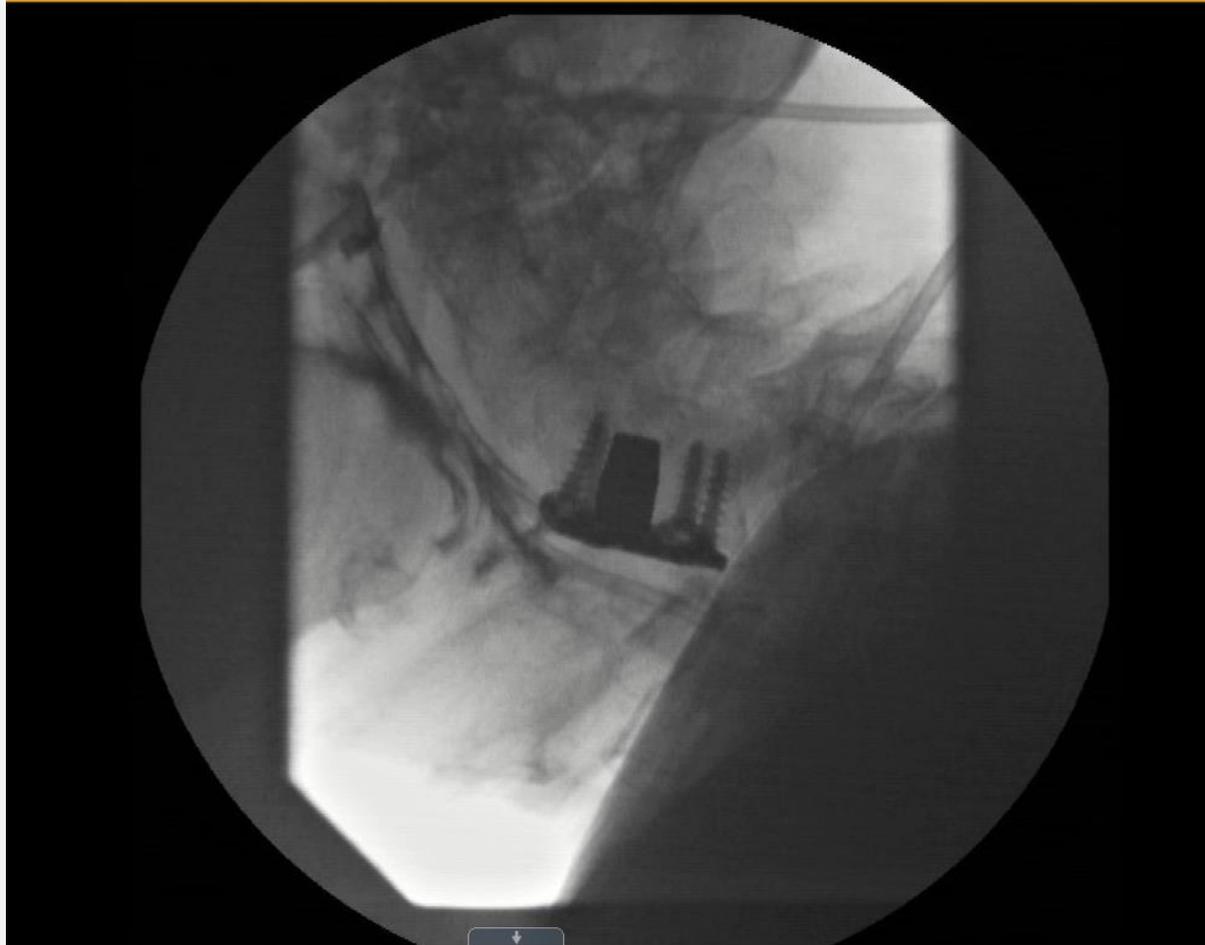
VFSS-THICK FROM A SPOON



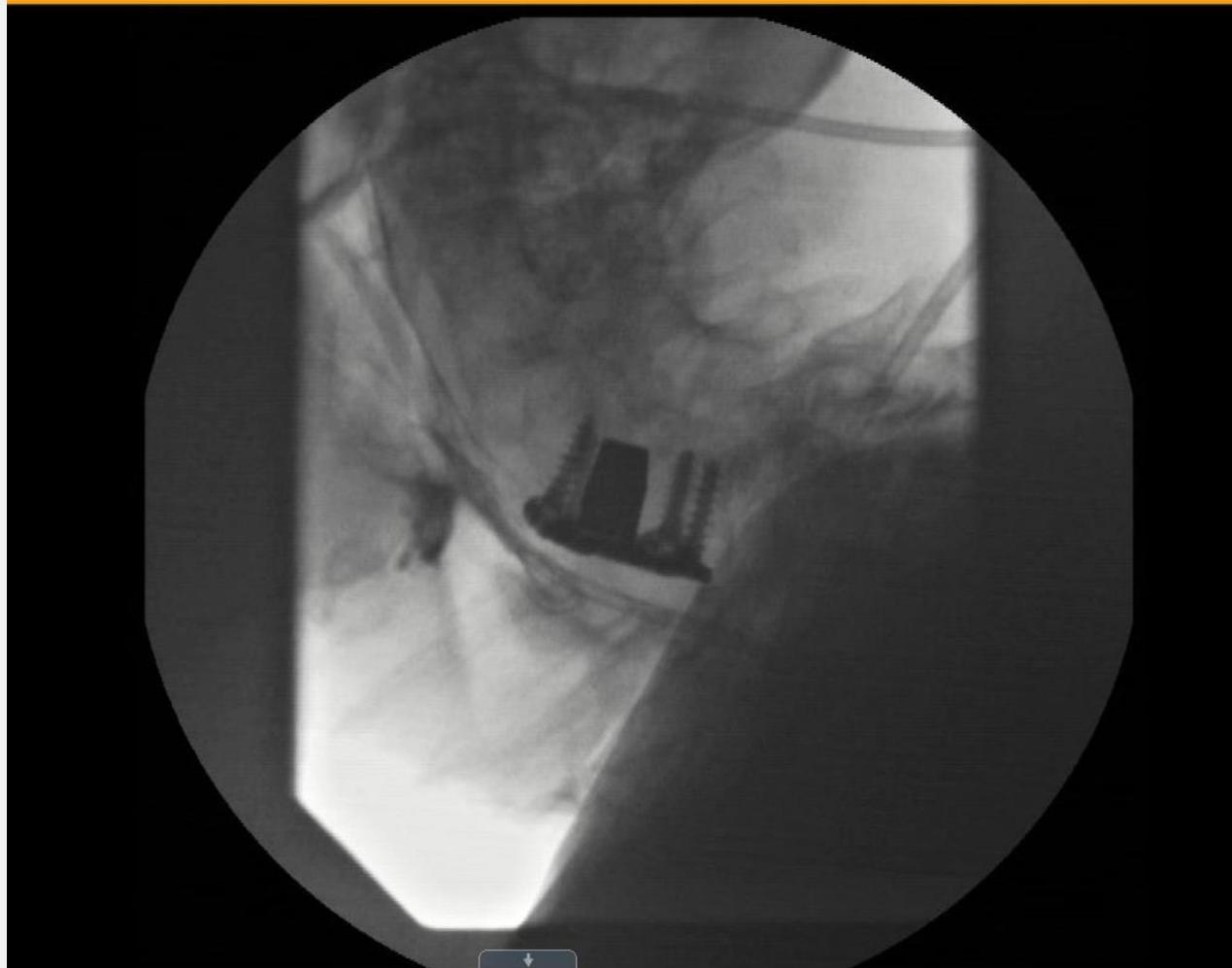
VFSS FOLLOWING IVIG-THIN BARIUM



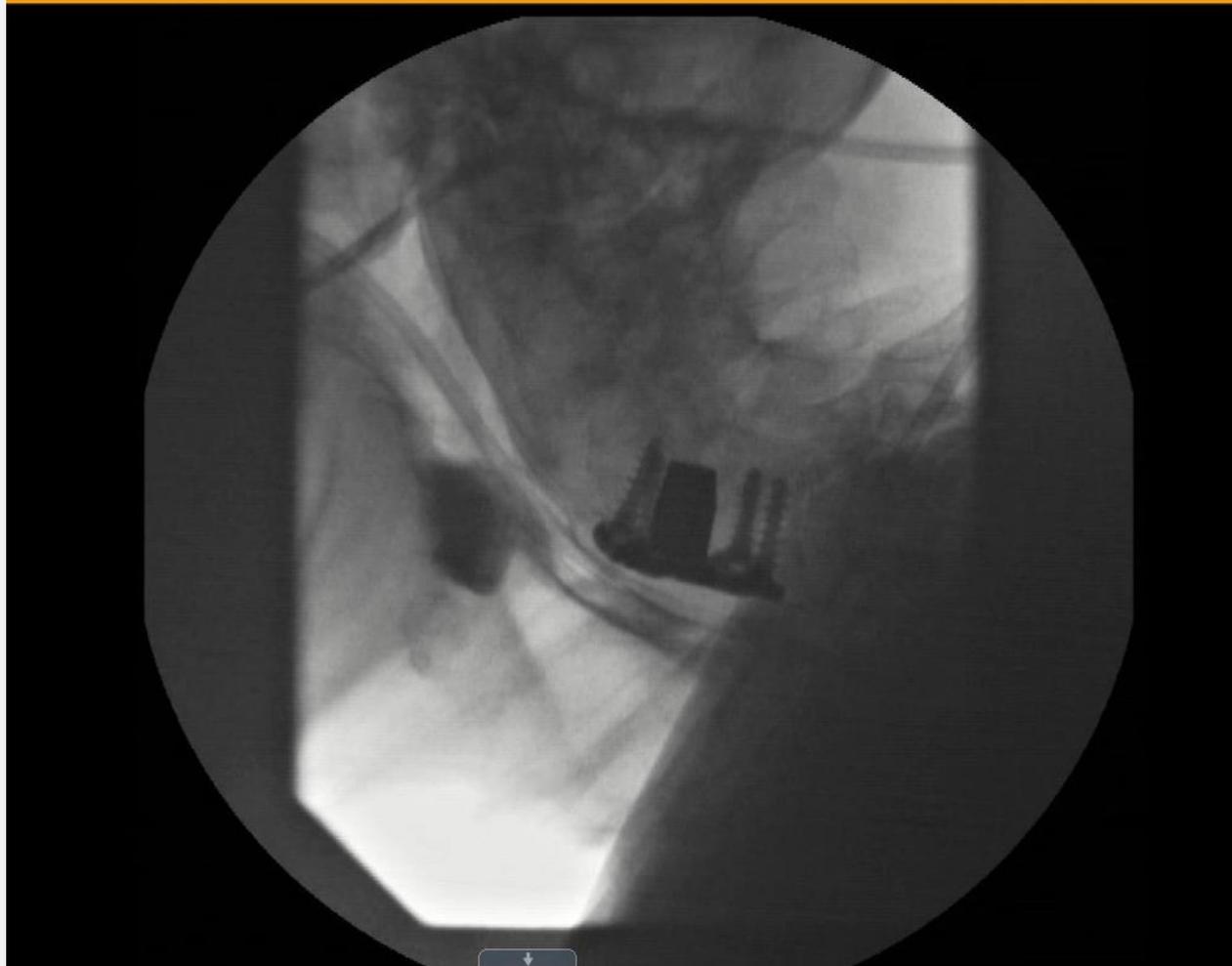
VFSS FOLLOWING IVIG-THICK BARIUM



VFSS FOLLOWING IVIG-PUDDING



VFSS FOLLOWING IVIG-FRUIT



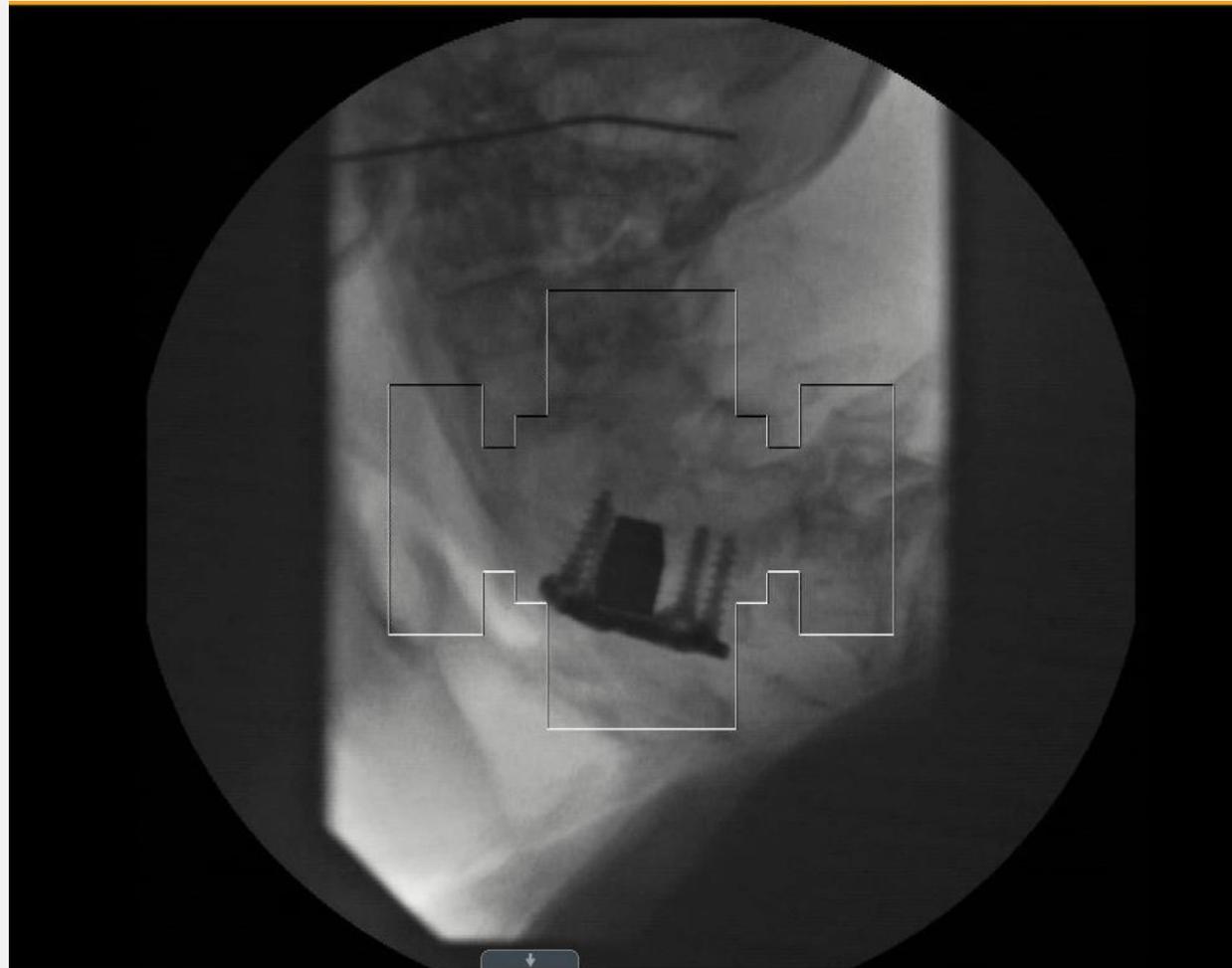
VFSS WHEN NOT IN FLARE THIN LIQUID



VFSS WHEN NOT IN FLARE PUREE



VFSS WHEN NOT IN FLARE COOKIE



CASE STUDIES

- Case study #3
- 68 year old woman with PMH of HTN, DM and Myasthenia who was admitted after being found lying unresponsive at home

CASE STUDIES

- **Oral Motor Examination:**
Facial/labial: Symmetric at rest and with activation. Fair labial ROM for pucker/smile.
Diminished strength on resistance.
Mandibular: Reduced ROM for opening/lateralization. Diminished strength to resistance.
Lingual: Protrusion midline; slow alternating movements. Diminished strength to resistance.
Noted to fatigue.
Palatal: Reduced elevation bilaterally.
- **Motor Speech Perceptual Examination:**
Phonation/Voice: Hypophonic and breathy.
Articulation: Reduced precision. Noted to be fatigable with use.
Resonance: Hypernasal.
Prosody: Short utterances. Lack of use of stress/inflection.
Speech intelligibility was judged to be 75%.

CASE STUDIES

- **Oral Agility:** Diadochokinetic rates were obtained for five second trials:

Test	Patient Performance	Norm
/pa/	22	28
/ta/	18	28
/ka/	14	25
/pataka/	6	8-10

CASE STUDIES

- **Swallowing:**
Trials included:
Ice chips
Thin liquid (water) via spoon, cup, and straw
Puree (applesauce)
Further trials deferred due to concern for aspiration.
- Performance notable for:
Reduced labial seal on spoon and cup.
Reduced ROM for mastication of ice chips; overall, slow to form/clear oral bolus.
No significant residue noted within the oral cavity post swallow.
Multiple swallows for water and puree boluses.
Immediate weak cough and throat clear following trials of purees.

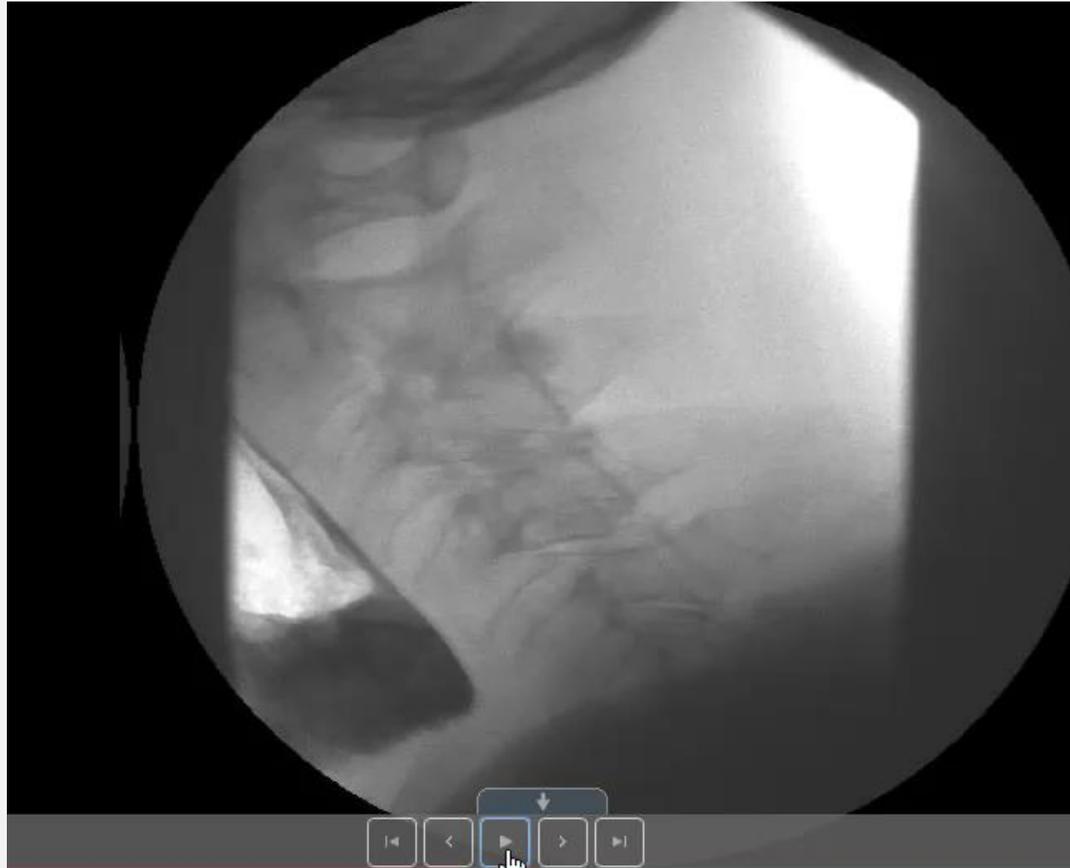
CASE 3: THIN VIA SPOON



PUDDING



WATER WASH



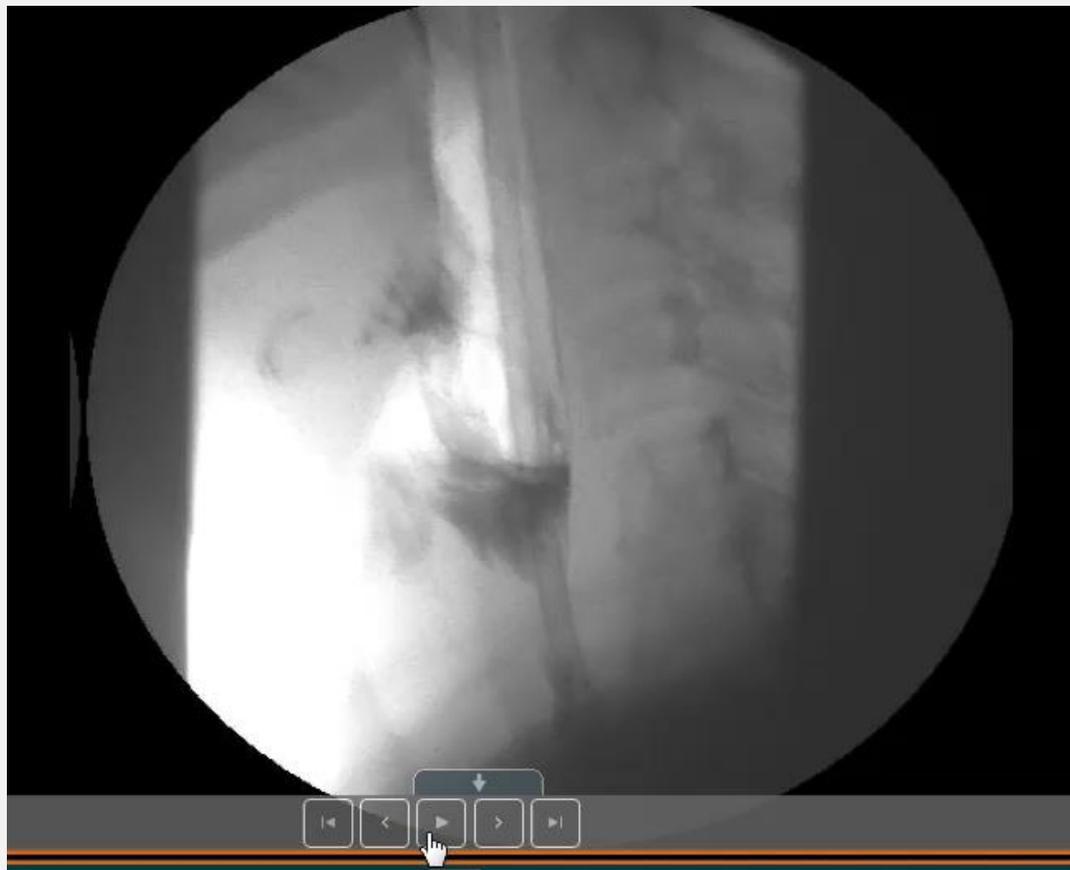
CASE 3 REPEAT: THIN VIA SPOON



THIN VIA CUP



THICK VIA CUP



PUDDING



WATER WASH WITH HEAD TURN



PUDDING WITH R HEAD TURN



THIN LIQUID A-P



CASE STUDIES

- Case study #4
- 69-year-old woman who presented to the ED with a Myasthenia Gravis exacerbation
- PMH:
 - CKD, HTN, DM2
- On a regular diet prior to admission

CASE STUDIES

- **Oral Motor Examination:**
Facial/labial: Fair labial ROM for pucker/smile. Reduced strength to resistance. Unable to create labial seal to fill her cheeks with air.
Mandibular: Fair ROM for opening/lateralization. Fair strength to resistance. On stress testing, noted to have decreased strength.
Lingual: Protrusion midline. Reduced ROM and strength to resistance. On stress testing, noted to have decreased strength.
Palatal: Reduced elevation bilaterally. Reduced gag.
- **Motor Speech Perceptual Examination:**
Respiratory support for speech: Sustained phonation of /a/: 10 seconds (norm is 16-20). Reduced breath support for conversational speech.
Phonation/Voice: Hoarse and breathy.
Articulation: Moderately reduced precision related to oral motor weakness. Noted to fatigue with stress testing.
Resonance: Moderate hypernasality.
Prosody: Intact prosody.
Speech intelligibility was judged to be 100%.

CASE STUDIES

- **Oral Agility:** Diadochokinetic rates were obtained for five second trials. Disturbances noted in the /ka/ sound due to reduced palatal and lingual range of motion and strength

Test	Patient Score	Norm
/pa/	18	28
/ta/	17	28
/ka/	13	25
/pataka/	5	8-10

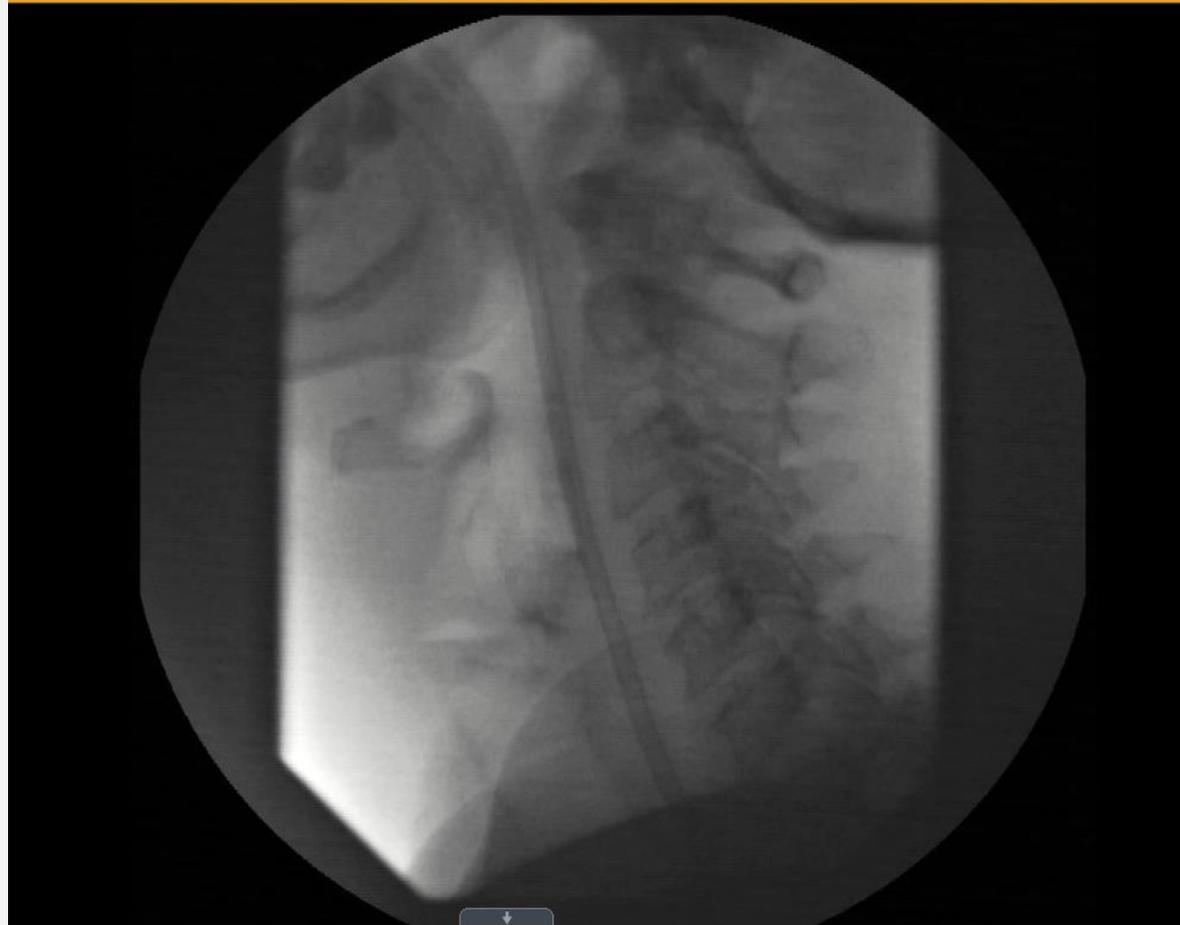
CASE STUDIES

- **Swallowing:**
Trials included:
Ice chips
Thin liquid (water) via cup
Puree (applesauce)
- Performance notable for:
Fair labial seal; no anterior spill.
Mastication was functional for ice chips.
Swallow onset appeared timely
Multiple swallows for liquid and pureed boluses.
No significant oral residue
On puree, patient had weak cough and throat clear, required multiple ice chips to wash down reported residue.

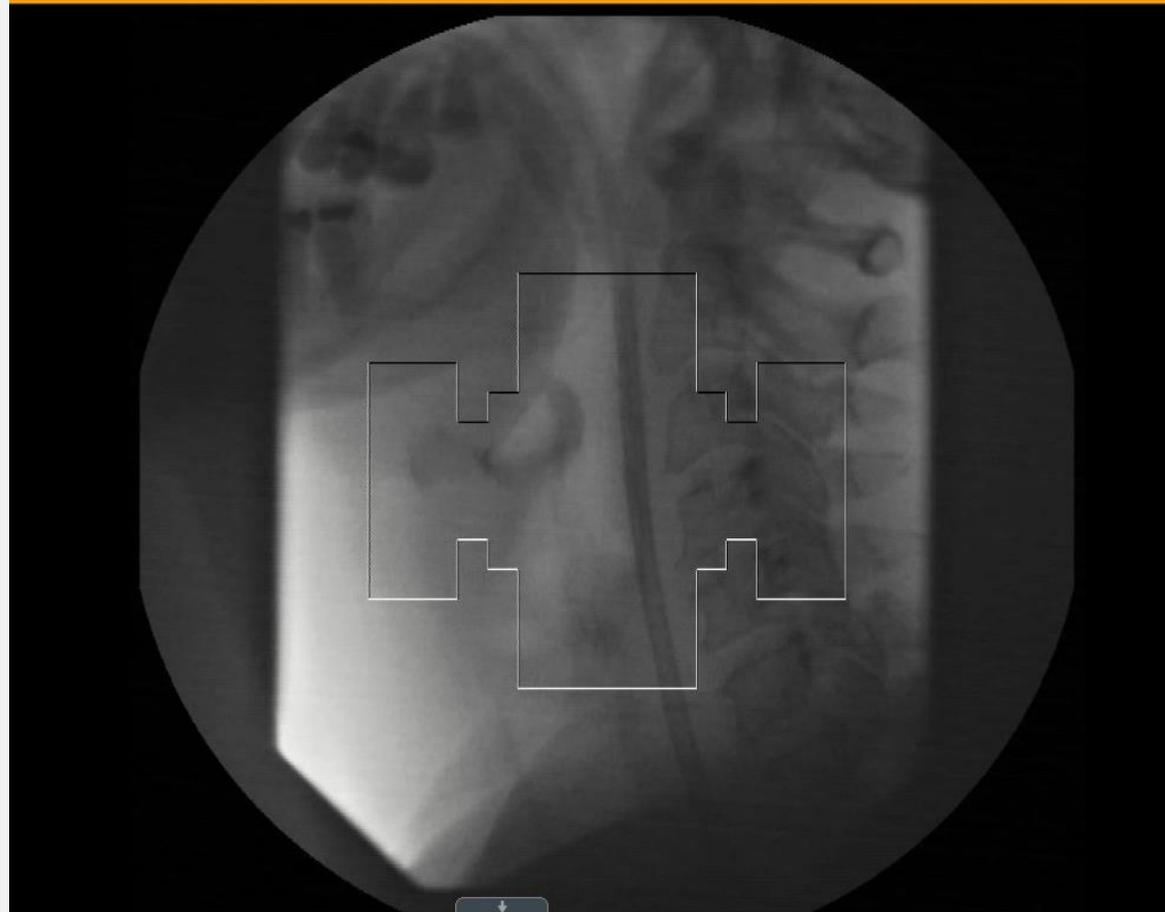
VFSS FOLLOWING IVIG THIN LIQUID



VFSS FOLLOWING IVIG THICK LIQUID



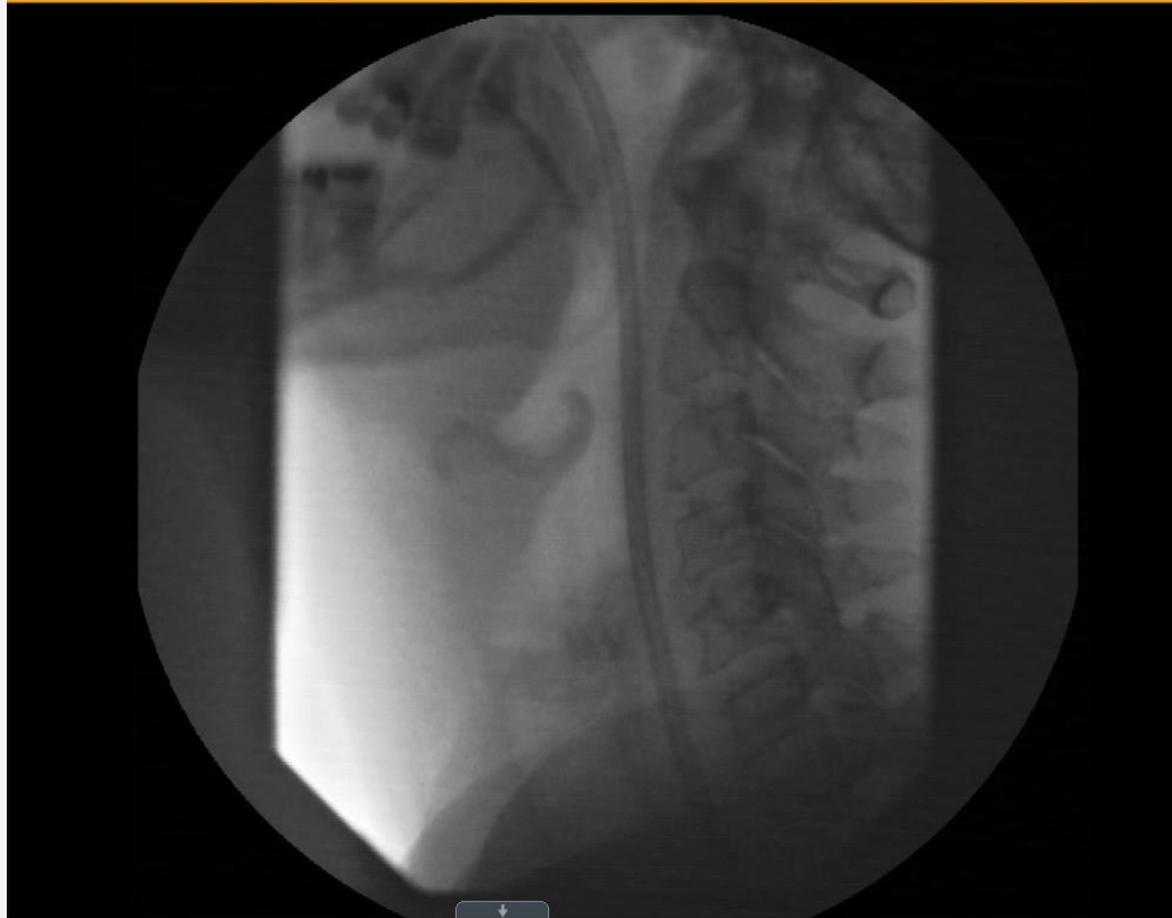
VFSS FOLLOWING IVIG PUREE



VFSS FOLLOWING IVIG DUAL CONSISTENCY



VFSS FOLLOWING IVIG SOLID



VFSS FOLLOWING IVIG AP-THICK LIQUID



CASE STUDIES

- Case study #5
- 77 year-old man with past medical history of refractory, acetylcholine receptor positive myasthenia gravis who presented 5 day history of dysphagia and dysarthria at home.
- He currently receives IVIG treatments every other week as an outpatient

CASE STUDIES

-
- **Oral Motor Examination:**
Facial/labial: Fair labial ROM for pucker/smile. Reduced strength to resistance.
Mandibular: Fair ROM for opening/lateralization. Reduced strength to resistance.
Lingual: Protrusion midline. Fair ROM and reduced strength to resistance.
Palatal: Mildly reduced elevation bilaterally. Reduced gag.
-
- **Motor Speech Perceptual Examination:**
Respiratory support for speech: Sustained phonation of /a/: 14 seconds (norm is 16-20).
Phonation/Voice: Mildly hoarse, hypophonic.
Articulation: Trace imprecision.
Resonance: Trace hypernasality.
Prosody: Unremarkable.
Speech intelligibility was judged to be 100%.

CASE STUDIES

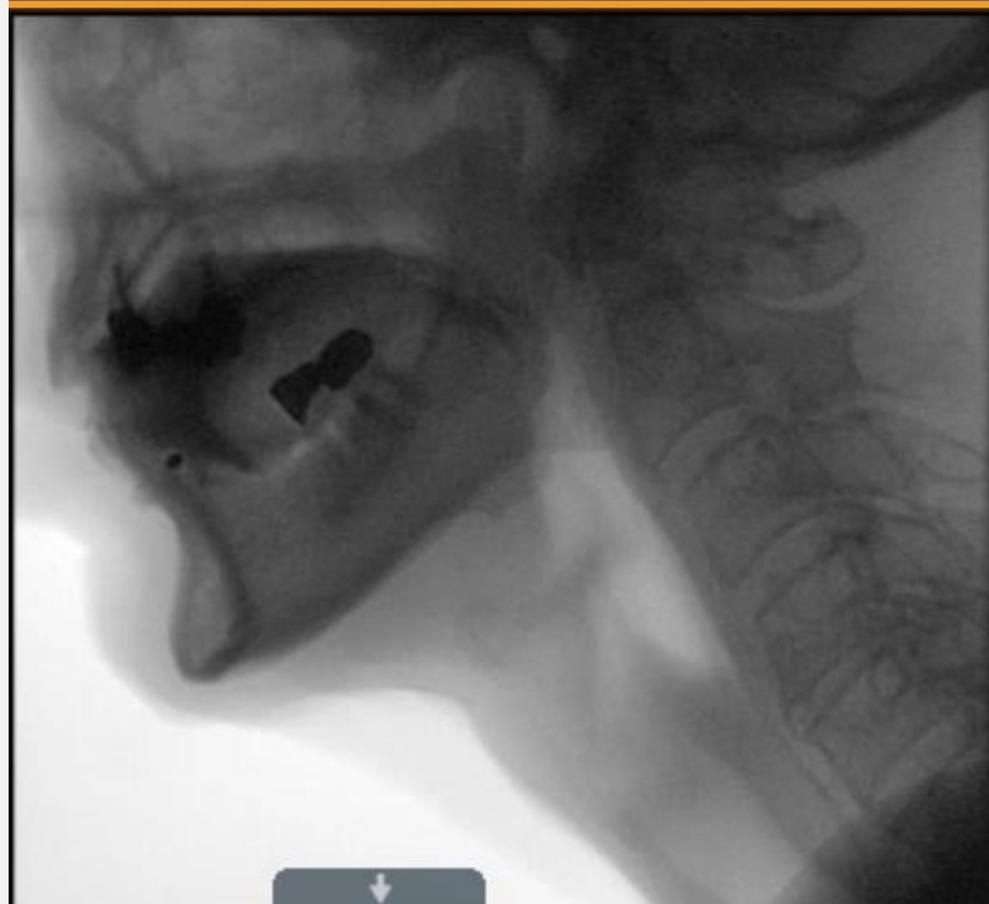
- **Oral Agility:** Diadochokinetic rates were obtained for five second trials. Disturbances noted in the /ka/ sound due to reduced palatal and lingual range of motion and strength

Test	Patient Score	Norm
/pa/	18	28
/ta/	20	28
/ka/	17	25
/pataka/	3	8-10

CASE STUDIES

- **Swallowing:**
Trials included:
Ice chips
Thin liquid (water) via spoon, cup, and straw
Puree (applesauce)
Dual consistency (fruit cocktail)
Solid (Cookie)
-
- Performance notable for:
Fair labial seal; no anterior spill.
Fair ROM for mastication. Mastication was slightly prolonged, but functional.
Swallow onset appeared timely.
Intermittent excess swallows.
No significant oral residue.
No cough, throat clear or change in vocal quality.

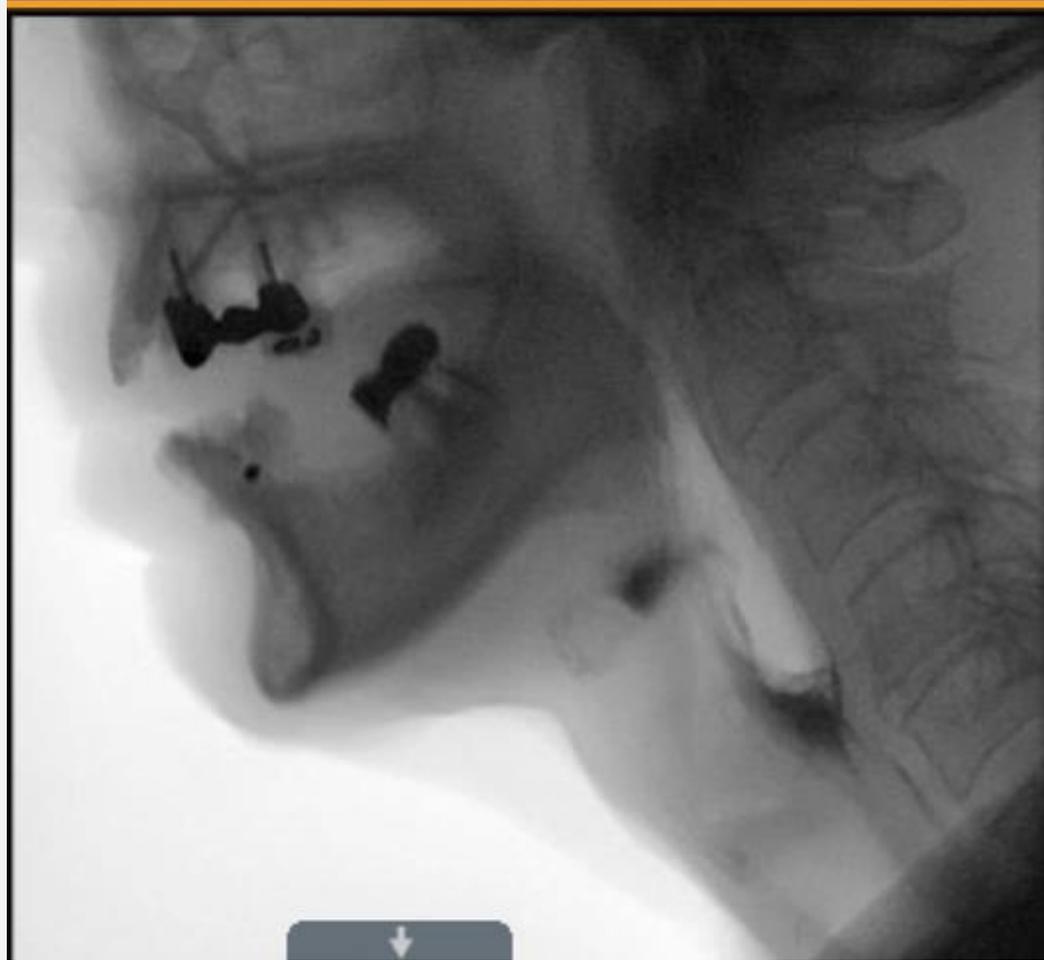
VFSS THIN LIQUID VIA CUP



VFSS THICK LIQUID VIA CUP



VFSS WATER VIA CUP



VFSS PUDDING VIA CUP



VFSS THIN LIQUID WASH



CONCLUSION

- A comprehensive motor speech evaluation is critical in evaluation of patients with Myasthenia Gravis.
- Understanding the primary physicians plan for medical intervention is critical to help with the speech-language pathology treatment planning.
- Formal imaging is important in this population to help guide the treatment plan
- Exercise and electrical stimulation are contraindicated in patients with Myasthenia Gravis.

REFERENCES

- Conti-Fine, B. M., Milani, M., & Kaminski, H. J. (2006). Myasthenia gravis: past, present, and future. *The Journal of Clinical Investigation*.
- Fda Approves Soliris For Particular Type Of Myasthenia Gravis
- Margarida Azevedo - <https://myastheniagravisnews.com/2017/10/25/fda-approves-soliris-for-a-particular-type-of-myasthenia-gravis/>
- Fowler, S. B., Herrington, J. B., Koopman, W. J., & Ricci, M. (2013). Care of the Patient with Myasthenia Gravis: AANN Clinical Practice Guideline Series. *American Association of Neuroscience Nurses*, 1-32.
- Higo, R., Nito, T., & Tayama, N. (2005). Videofluoroscopic Assessment of Swallowing Function in Patients with Myasthenia Gravis. *Journal of the Neurological Sciences* , 231, 45-48.

REFERENCES

- Kluin, K. J., Bromberg, M. B., Feldman, E. L., & Simmons, Z. (1996). Dysphagia in Elderly Men with Myasthenia Gravis. *Journal of the Neurological Sciences* , 138, 49-52.
- Ludlow, C. L., (2008). Electrical Stimulation and Dysphagia: What We Do and Don't Know. *The ASHA Leader*, 13, 8-11.
- Mestinon (pyridostigmine): Uses, Dosage, Side Effects, Interactions, Warning
<https://www.rxlist.com/mestinon-drug.htm#warnings>
- Myasthenia Gravis
<https://www.mayoclinic.org/diseases-conditions/myasthenia-gravis/diagnosis-treatment/drc-20352040>
- Rison, R. A. (2009). Reversible Oropharyngeal Dysphagia Secondary to Cricopharyngeal Sphincter Achalasia in a Patient with Myasthenia Gravis: A Case Report. *Cases Journal* , 1-3.
- Rituximab For Myasthenia Gravis
Maria Almeida - <https://myastheniagravisnews.com/rituximab-for-myasthenia-gravis/>