Road to Recovery: Dysphagia Treatment With An Adult with SMA Type II

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Objective

Spinal Muscular Atrophy (SMA) is a genetic and degenerative motor neuron disease that affects muscle control including the muscles responsible for head and neck control and swallowing (Kesting, n. d.). There is limited research on the benefits of exercise as a therapeutic modality for people with Spinal Muscular Atrophy (SMA) (Swoboda et al., 2007) and no existing research specifically targeting pharyngeal or laryngeal strengthening to improve swallow function. As demonstrated by the positive outcome in this case study, speech-language pathologists should be aware that various modalities for dysphagia treatment such as biofeedback, neuromuscular electrical stimulation (NMES), and an exercise based approach in conjunction with compensatory strategies may be effective in the swallowing rehabilitation of patients with patients with SMA.

Timeline of Events

- Prior to January 2014 patient on regular diet/ thin liquids
- 1/9/14 Admitted to hospital with peritonitis and pneumonia.
- February 2014 - Trach and ventilator dependent. Required PEG tube for nutrition.
- 3/10/14 patient de-cannulated.
- 3/20/14 MBS with silent aspiration, non-functional swallow.
- 3/25/15 Patient discharged. Taking some PO against advice.
- May 2014 Referred to outpatient SLP by MD.
- 5/19/14 Outpatient SLP Evaluation revealed no hyolaryngeal movement on palpation.
- 5/27/14-9/5/14 Attended dysphagia therapy 2x per week.
- Patient attended total of 18 therapy sessions.
- 8/16/14 Repeat MBS. No aspiration on any consistency.
- Patient reports he is doing well. Deferred repeat barium swallow at 1 year follow-up.

Case Summary

The patient is a 27 year old male with a childhood diagnosis of SMA Type II. He had been a known chronic, silent aspirator; however, he maintained a PO diet without respiratory issues or pneumonia. In January of 2014, he was hospitalized with peritonitis and pneumonia. His hospital course was complicated by tracheostomy and ventilator dependency for respiratory failure as well as PEG tube placement for primary nutrition. He remained NPO during his hospital stay deconditioning. A repeat modified barium swallow (MBS) was completed on 3/20/14, revealing a severe oral-pharyngeal dysphagia marked by decreased tongue base retraction, delayed swallow, atonic pharynx without evident contraction, absent epiglottic inversion and no laryngeal sensation resulting in aspiration of all consistencies. In May 2014, thewhich resulted in worsening of pre-existing dysphagia due to

Case Summary (continued)

patient began dysphagia therapy with treatment modalities including Hi-Volt NMES utilizing single channel- suprahyoid electrode placement , and biofeedback. NMES was paired with PO trials, base of tongue and pharyngeal strengthening exercises, and compensatory strategy training. In addition to participating in therapy twice weekly for a total of 18 visits, the patient was compliant with an aggressive home exercise program. A repeat modified barium swallow on 8/26/14 demonstrated significant improvement in oral-pharyngeal swallow function marked by improved epiglottic movement and pharyngeal contraction. With the use of a supraglottic swallow, the patient had only intermittent penetration and no episodes of aspiration on all consistencies.

Treatment Outcomes

Prior to onset of therapy, the patient was aspirating all consistencies as evidenced by performance on a pre-treatment MBS. The Eating Assessment Tool (EAT-10) and the Functional Oral Intake Scale (FOIS) were administered for patient rating of performance, and ASHA National Outcome Measures (NOMS) were rated by the clinician both pre- and post-treatment with outcomes as follows:

Pre-Treatment Measures:	**Post-Treatment Measures:**
EAT-10: 10/40	EAT-10: 5/40
FOIS: 5	FOIS: 6
ASHA NOMS: 3	AHSA NOMS: 5

At time of discharge, the patient had no evidence of aspiration of any consistency on post-treatment MBS. The patient was consuming a soft solid diet and thin liquids. His PEG tube was removed.

Conclusions

There is a lack of evidence in the literature to support exercise based therapy and the use of NMES for dysphagia rehabilitation in patients with SMA. In this case study, an intense exercise-based therapy program including the modality of Hi-Volt NMES, biofeedback, and compensatory strategy training, resulted in significant improvement of his premorbid swallow function. Pre-treatment aspiration risk, that had worsened in severity due to deconditioning as a result of a prolonged hospital stay and NPO status, was remediated. SLPs treating children and adults with SMA should consider these patients potential candidates for an exercise based treatment program to either maintain current function or for the purpose of rehabilitation of function. In addition, there may be a role for non-traditional treatment modalities such as NMES, progressive lingual resistance exercise, and biofeedback. As dysphagia specialists, we must continue to expand the research base for this patient population.

References

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Additional Resources

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