

# The effects of therapeutic intervention on a case of dermatomyositis

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## Background

Dermatomyositis is an acquired systemic inflammatory condition affecting the striated muscles.

It is characterised by proximal weakness, muscle tenderness, fatigue and a characteristic skin rash.

Dysphagia is a known complication of this condition, occurring in 10-73% of patients, and it primarily affects the skeletal muscle-activated oropharyngeal phase of swallowing (Kwon et al., 2018). There is limited research supporting the benefits of exercise-based dysphagia

therapy in patients with dermatomyositis, as well as the recommended timing of intervention, although recent evidence suggests the importance of commencing treatment regimens early in the disease process, in order to reduce inflammation, fatigue and build muscle (Okogbaa & Batiste, 2019).

The following case study demonstrates the positive outcomes of early implementation of an intensive dysphagia therapy programme in a patient with dermatomyositis.

## Case Presentation

The patient is a 59 year-old female who was admitted to acute hospital with worsening muscle weakness, dysphagia, aspiration pneumonia and reduced FVC on the 1<sup>st</sup> of March 2019.

In ITU she was treated with high dose steroids and antibiotics. A nasogastric tube (NGT) was inserted to meet her nutrition and hydration needs. Due to significant and diffuse oedema, insertion of the NGT was problematic, which contributed to delays in establishing early nutrition. As a result, blood sugar levels rose as they were challenging to manage and were likely exacerbated by this inflammatory state. She was reviewed by rheumatology, who weaned her down steroids and started her on methotrexate.

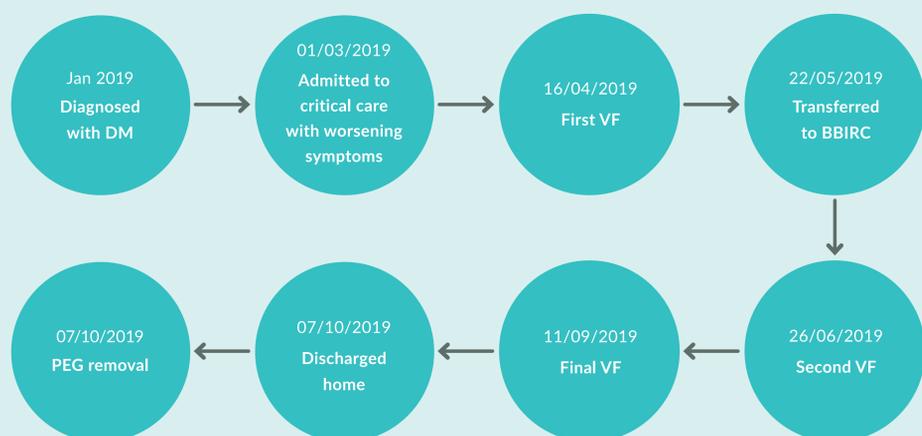
In close discussion with the rheumatology team and the neurologist no swallowing intervention was implemented in the early stages of the disease due to the extent of the inflammatory process. Furthermore, completion of a videofluoroscopy (VF) swallow study (VFSS) was necessary to inform choice of exercises and to rule out or identify suspected silent aspiration prior to implementation of a tailored therapy programme.

Following homeostasis of the condition, a VF was conducted on the 16<sup>th</sup> of April, which revealed severe oropharyngeal dysphagia with risk of silent aspiration. Specific impairments included: significantly reduced base-of-tongue to pharyngeal wall contact, incomplete swallow with reduced hyoid excursion and no epiglottic deflection, significant pharyngeal residue with inconsistent cough response, and minimal transfer of bolus through the upper oesophageal sphincter.

It was recommended that the patient remain Nil By Mouth (NBM) and that she commence modified Shaker and effortful swallow exercises, as well as strict oral trials only under supervision of SALT as part of dysphagia therapy. A percutaneous endoscopic gastrostomy (PEG) was subsequently inserted.

On the 22<sup>nd</sup> of May she was admitted to Blackheath Brain Injury Rehabilitation Centre for ongoing intensive rehabilitation. An intensive dysphagia therapy programme was commenced, which included intensive daily therapy with modified Shaker, Masako, Mendelssohn and effortful swallow. Interprofessional goals were integral to her recovery including increasing independence and self-control at mealtimes.

## Timeline of Events



## Treatment Outcomes

Prior to onset of therapy, the patient was not tolerating an oral diet and had been found to be at high risk of aspiration during an early VF.

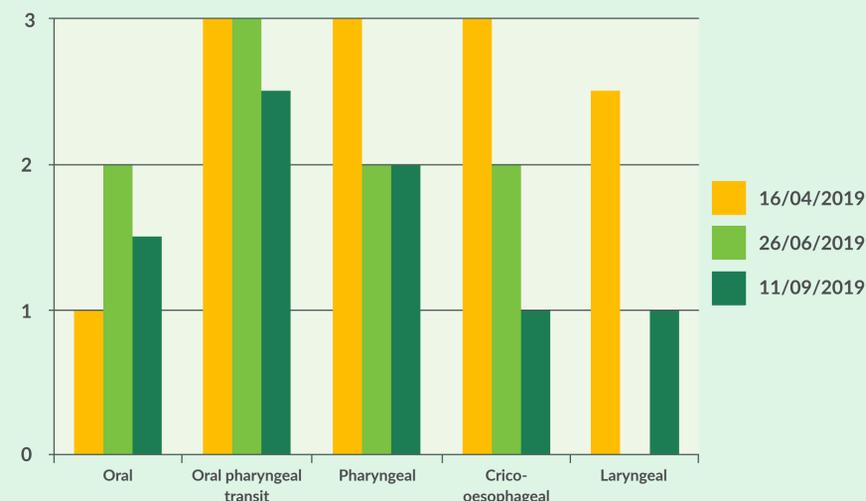
On bedside examination she presented with prolonged bolus formation and delayed A-P transport; delayed pharyngeal swallow trigger (~4 seconds) and significantly reduced hyolaryngeal elevation/excursion on palpation. Occasional throat clears and multiple re-swallows (~4

per bolus) were observed. The patient reported excellent sensation and was able to state when the bolus had been cleared from her pharynx. Objective and subjective outcome measurements were taken over the course of the patient's rehabilitation pathway; these included the New Zealand Index for Multidisciplinary Evaluation of Swallowing (NZIMES) and the Australian Therapy Outcome Measures (AusTOMs). Outcomes are as follows:

### Australian Therapy Outcome Measures (AusTOMs)

AusTOMs Therapy Outcome Measures: Swallowing	Impairment	Activity Limitation	Participation Restriction	Distress/Wellbeing
Admission: 05/2019	2	1	2	2
Review: 09/2019	3	4	4	4
Discharge: 10/2019	5	5	5	5

### New Zealand Index for Multidisciplinary Evaluation of Swallowing (NZIMES)



Final VF revealed more timely A-P transport of the bolus, improved base-of-tongue to pharyngeal wall contact, pharyngeal swallow trigger timeliness and epiglottic deflection.

At the time of discharge the patient was tolerating a Level 7 (regular) consistency diet and Level 0 (thin) fluids, and was no longer requiring enteral nutrition. Her PEG tube was removed shortly post discharge.

## Conclusions

There is limited evidence-based research to support the early implementation of dysphagia rehabilitation in patients with dermatomyositis.

It was a commonly held belief that dysphagia in patients with dermatomyositis occurred as a result of an esophageal disorder, particularly failed UES relaxation. However recent research shows that it is more likely due to impaired pharyngeal muscle contraction, stemming from weakness in the suprahyoid muscles. The limited

literature also suggests that as well as an exercise-based approach, non-traditional treatment modalities such as neuromuscular electrical stimulation (NMES) would provide positive outcomes. Due to a lack of resources, NMES could not be implemented in this case; however, **an intensive oropharyngeal dysphagia therapy programme in the form of daily repetitions of the Masako, Mendelssohn, modified Shaker and effortful swallow exercises, resulted in significant improvements in the patient's swallow function.**