eNeurologicalSci 20 (2020) 100255

Contents lists available at ScienceDirect

eNeurologicalSci

journal homepage: www.elsevier.com/locate/ensci

Case report

Therapy-resistant dysphagia successfully treated using pharyngeal electrical stimulation in a patient with the pharyngeal-cervical-brachial variant of the Guillain-Barré syndrome

Sebastian Beirer*, Wolfgang Grisold, Jennifer Dreisbach

Sozialmedizinisches Zentrum Süd - Kaiser-Franz-Josef-Spital mit Gottfried von Preyer'schem Kinderspital, Kundratstraße 3, 1100, Vienna, Austria

ARTICLE INFO

Keywords:
Guillain-Barré syndrome
Dysphagia
Pharyngeal electrical stimulation
Decannulation
Single case

ABSTRACT

Pharyngeal electrical stimulation (PES) is a novel treatment for oropharyngeal dysphagia resulting from neurogenic causes such as stroke, prolonged intubation, tracheostomy, or multiple sclerosis, and may be effective in other medical conditions such as Guillain-Barré syndrome (GBS). A 74-year-old male patient with a pharyngeal-cervical-brachial (PCB) variant of GBS, who had been tracheotomised due to severe and persistent swallowing difficulties (dysphagia) unresponsive to traditional speech and language therapy, received PES therapy as a final treatment option. Swallow performance evaluated before and after PES using fibreoptic endoscopic evaluation of swallowing, videofluoroscopy and clinical bedside assessment, showed significant improvement in swallowing resulting in safe tracheostomy decannulation 18 days after PES.

In PCB GBS, we present the potential benefit of PES for the treatment of persistent dysphagia and faster tracheostomy decannulation. PES was safe and may be beneficial in other neurologic disorders, where traditional dysphagia therapies have proved unsuccessful.

1. Introduction

Dysphagia, or "disordered swallowing", can seriously compromise nutritional status, lead to significant comorbidity, and increase risk of death. Dysphagia can be associated with numerous neuromuscular diseases and present different mechanisms, such as loss of motor neurons, damage to caudal brain nerves (neuropathies or local nerve lesions), neuromuscular transmission or musculoskeletal disorders [1]. In inflammatory neuropathies, such as Guillain-Barré syndrome (GBS) and variants, dysphagia is rare but increasingly described in the literature, with severe cases requiring intensive care treatment [2–4]. GBS is an immune-mediated polyneuropathy characterized by rapidly progressive weakness and sensory loss. In the pharyngeal-cervical-brachial (PCB) variant, a preferred involvement of the caudal cranial nerves results in dysphagia, though the exact pathomechanism for dysfunction is still unclear. It might involve axonal demyelination, conduction block, or combinations.

Traditional speech and language therapies (SLT) for dysphagia, typically consist of bolus modifications, compensatory strategies and muscle strengthening exercises but have limited efficacy [5]. Pharyngeal electrical stimulation (PES) is a novel, simple and safe technique for restoring neurological control of swallowing in the brain. PES has been shown to be effective in enabling faster decannulation of severe stroke patients compared to sham in both pilot [6] and randomised controlled [7] studies; and reducing the prevalence of pneumonia and frequency of reintubation in orally intubated intensive care unit patients [8].

We present the results of PES for the treatment of traditional therapy-resistant severe dysphagia in a patient with the PCB variant of GBS.

2. Materials, procedures, and patient

Written consent allowing data collection and publication was obtained from the patient.

Swallowing was evaluated using fibreoptic endoscopic evaluation of swallowing (FEES), videofluoroscopy and clinical bedside assessments. The Gugging Swallowing Screen (GUSS) score, consisting of indirect (assessment of vigilance, voluntary cough, throat clearing, and saliva swallowing) and direct swallows tests (sequential subtests of semisolid, liquid and solid textures) yielding a total score from 0 to 20 (0 = complete pathological swallow, high aspiration risk, 20 = normal swallow

* Corresponding author.
E-mail addresses: sebastian.beirer@gesundheitsverbund.at (S. Beirer), grisoldw@gmail.com (W. Grisold), jennifer.dreisbach@gesundheitsverbund.at (J. Dreisbach).

https://doi.org/10.1016/j.ensci.2020.100255
Received 21 April 2020; Received in revised form 27 June 2020; Accepted 10 July 2020
Available online 15 July 2020

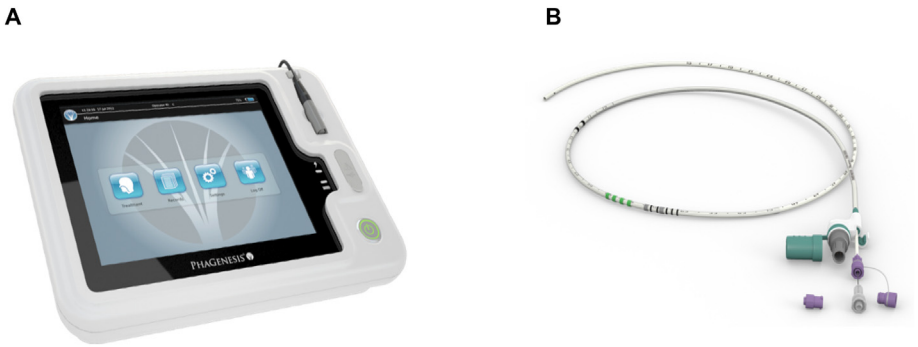

Fig. 1. Phagenyx® Pharyngeal Electrical Stimulation, a medical device comprising a base station with a touch screen user interface (A) and a sterile single-patient use catheter (B) that can be used to deliver nutrition and hydration for up to 30 days after insertion.

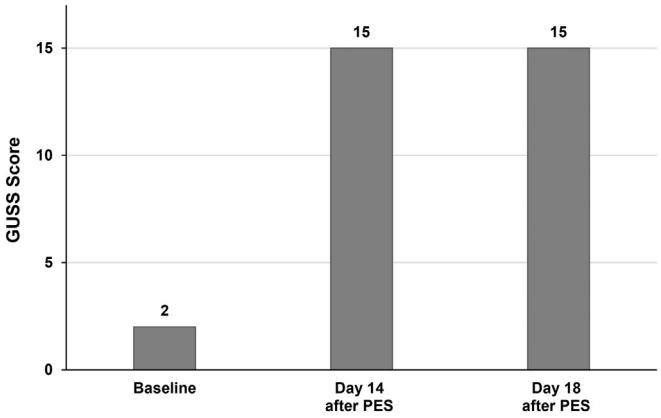

Fig. 2. Schematic timeline of events and outcomes from hospital admission until recovery after PES treatment.

without aspiration risk) [9], was used to determine dysphagia severity level before (baseline) and at several timepoints after PES. Since solids were not tested, the maximum score achievable was 15.

PES was delivered using the Phagenyx® system (Phagenesis Ltd., Manchester, UK) (Fig. 1). This involved insertion of a specially designed, single-patient use, nasogastric catheter with built-in electrodes positioned in the oropharynx and used to deliver trains (200 μs pulses at 5 Hz) of PES for ten minutes per day for three consecutive days. Prior to each treatment session, the current intensity (mA) of PES was optimised by the operator in response to the patient's responses as reported previously [7,8].

2.1. Case presentation

2.1.1. Diagnosis.

A 74-year-old male patient hospitalized in Sozialmedizinisches Zentrum Süd – Kaiser-Franz-Josef-Spital mit Gottfried von Preyer'schem Kinderspital (Vienna, Austria) for rapidly progressive dysphagia. Voluntary and involuntary swallowing was affected, which, in addition impaired nutritional intake, also led to constant aspiration followed by pneumonia. In addition to ankylosing spondylitis and medically controlled hypertension, the patient had no other relevant comorbidities. Clinically, a proximal weakness in the extremities and areflexia were observed. Through edrophonium test and repetitive stimulation, a neuromuscular transmission disorder was excluded. Peripheral neurography indicated a predominant axonal damage as the nerve conduction velocities showed axonal stressed changes and loss of the F-waves.

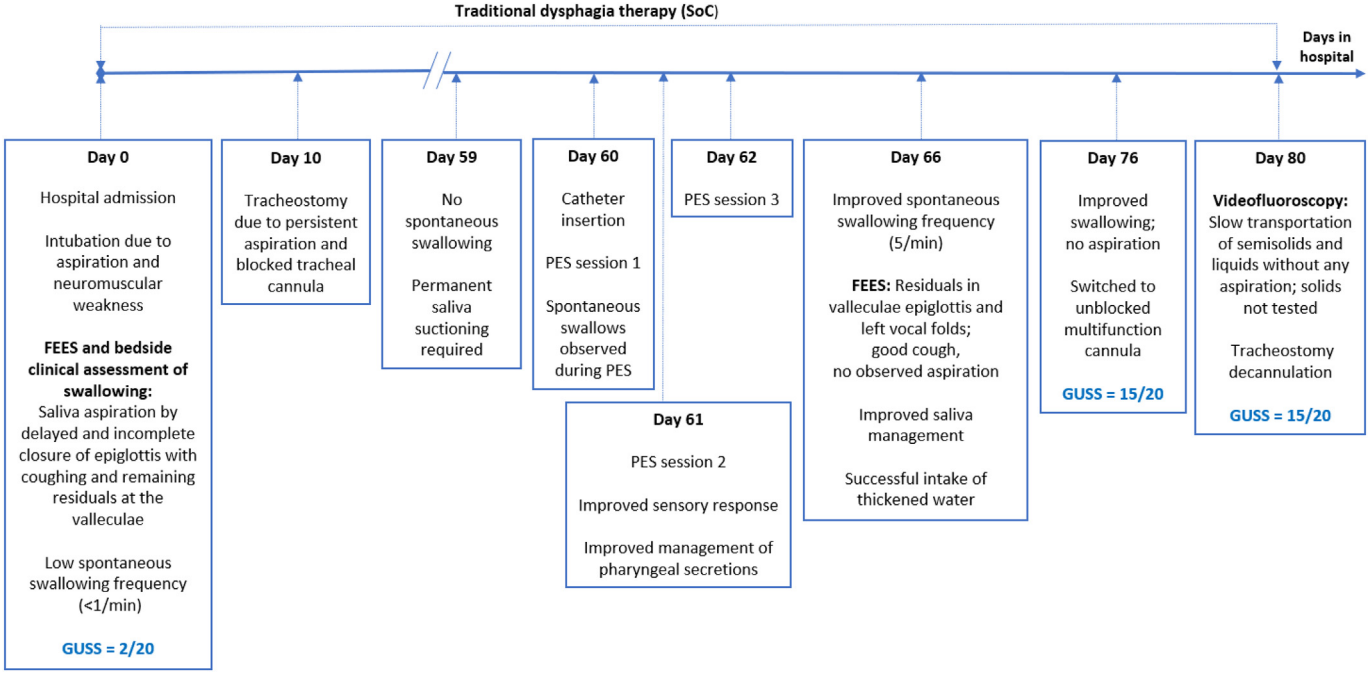

Fig. 3. Schematic timeline of events and outcomes from hospital admission until recovery after PES treatment

No further cranial nerve investigations were carried out. Based on electrophysiology, clinical signs and symptoms, a pharyngeal-cervical-brachial (PCB) variant of GBS was suspected and further confirmed serologically (GD1a, GQ1b, borderline detection of GD1b antibodies). Swallowing was significantly impaired and due to aspiration and neuromuscular weakness, the patient was rapidly intubated and ten days later, tracheotomized for persistent aspiration and blockage of the tracheal cannula. The patient required mechanical ventilation for 33 days and was fed exclusively by nasogastric tube in the first instance, and later through a percutaneous endoscopic gastrostomy tube. Therapeutically, a total of two series of intravenous immunoglobulin (IVIG) and a series of plasmapheresis were given.

2.2. Swallowing evaluation, SLT therapy and PES.

Baseline FEES showed incomplete epiglottic closure, complete paresis of both velum concomitant, coughing, residue in the valleculae and no pooling in the laryngeal vestibule. Swallowing frequency was less than 1/min with total saliva aspiration (GUSS 2/20). Dysphagia therapies including exercises to strength the tongue and improve range of motion, thermal and gustatory stimulation, and tracheostomy weaning over two months through the SLT department. Due to the patient's ankylosing spondylitis, pharyngeal muscle exercises, Mendelsohn manoeuvres, postural changes and other positional techniques were not feasible. Despite the SLT, there was no improvement in swallowing. As a final treatment option and because the patient repeatedly experienced saliva aspiration events due to tracheostomy decay, we opted to carry out an individual approach with PES. PES treatment was applied for 10-min daily for three consecutive days. Swallow performance was re-evaluated on day 14 and 18 post-stimulation. The patient continued to receive standard-of-care SLT.

2.3. Treatment outcome.

In the two months following admission and standard-of-care SLT, no swallow reactions had been observed. Remarkably, during the first PES session, the patient spontaneously swallowed approximately 40 times. The first stimulation-independent spontaneous swallowing acts occurred four days after PES and during FEES, there was minimal pooling of saliva at the valleculae with fine white out. Valleculae residue observed to drip onto the vocal folds caused coughing with good clearance of thickened water. Swallowing frequency had increased to 5/min with no aspiration. Further swallowing improvement was observed as clinical bedside assessment increased on Day 14 with GUSS of 15/20 and a stable state was achieved on Day 18 (GUSS of 15/20) (Fig. 2), so no further assessments were performed.

Considering the improved swallowing and lack of evidence of aspiration, the patient was switched to an unblocked multifunctional cannula two weeks after PES. Further, the patient was able to vocalise loudly and clearly indicating he was no longer aspirating heavily nor that the severe weakness of the respiratory muscles was still present. According to our hospital's standard guidelines, readiness for decannulation was confirmed and further supported by FEES and videofluoroscopy; thus, the tracheal cannula removed 18 days after last PES, or 80 days after hospital admission. Fig. 2 presents a summary timeline of events and outcomes. (See Fig. 3.)

After dysphagia recovery, as the patient still needed additional therapy for tetraparesis, hospital discharge occurred 70 days post-stimulation, after an overall hospitalisation length of 132 days. The patient was then transferred to a neurology rehabilitation centre for further mobilisation.

3. Discussion

We reported the case of a patient requiring orotracheal intubation and tracheostomy due to a PCB confirmed variant of GBS leading to complete abolishment of swallowing. After two months of traditional SLT with no response, PES was offered as a final treatment option and led to a sustained restoration of deglutition and subsequently to tracheostomy decannulation within 18 days of the final PES session.

This patient's severe dysphagia was caused by impairment of the caudal cranial nerve function. Although no ancillary investigations were performed, the rapid improvement of swallowing following PES may be due to conduction block reversal, or a change in the stimulus threshold, or a temporary increase of salivary substance P levels [10–12] by the PES. This is supported by the observation that the tongue was not affected by atrophy, which would have been the case if axonal damage was present. Moreover, PES appears to have reactivated sensory feedback pathways which are known to be critical to safe and efficient swallowing; this was evidenced in our patient by an improved sensory perception (dysesthesia) in the pharynx after the second PES session, an increased spontaneous swallowing and a better pharyngeal secretion management observed post-PES.

There is no pharmacological treatment for oropharyngeal dysphagia and removal of the tracheal cannula in tracheotomized GBS patients with persistently impaired deglutition is challenging. Patients presenting impaired swallowing and needing long-term institutional care due to delayed recovery represent a significant economic burden. This is, to our knowledge, the first report of a PCB GBS patient showing a fast recovery of swallowing dysfunction following PES. PES is shown to be a safe and efficient treatment of dysphagia in patients with stroke [7,10,13], brain injury [14], multiple sclerosis [15] and now GBS. Extrapolations to other neuromuscular swallowing disorders cannot be generalised from our observation, but PES treatment should be considered in individual cases where severe and persistent dysphagia is present.

4. Conclusions

In this patient case report, PES was easy to implement, led to a rapid long-lasting effect, and seems to be a safe therapeutic option for GBS patients presenting severe swallowing dysfunction, as no treatment-related adverse events were observed. In absence of currently established standard treatments, PES is a welcome alternative to facilitate swallowing recovery of dysphagic patients.

List of abbreviations.

FEES Fibreoptic endoscopic evaluation of swallowing
GBS Guillain-Barré syndrome
GUSS Gugging Swallowing Screen
IVIG Intravenous immunoglobulin
PES Pharyngeal electrical stimulation

Acknowledgements

The authors would like to thank Florence Boulmé, PhD, for writing assistance.

Declaration of interests

SB received honoraria (speaker fee) from Pfizer in 2019. WG received honoraria for lectures from AMGEN. The remaining author has nothing to declare.

Authors' contributions

All authors contributed equally to this work; all of them reviewed and approved the final manuscript.

References

[1] N. Audag, C. Goubau, M. Toussaint, G. Reychler, Screening and evaluation tools of dysphagia in adults with neuromuscular diseases: a systematic review, Ther Adv

Chronic Dis 10 (2019) 2040622318821622.
[2] T. Mengi, Y. Secil, T.K. Incesu, S. Arici, Z.O. Akkiraz, N. Gurgor, et al., Guillain-Barre syndrome and swallowing dysfunction, J Clin Neurophysiol 34 (5) (2017) 393–399.
[3] J.B. Schroder, T. Marian, P. Muhle, I. Claus, C. Thomas, T. Ruck, et al., Intubation, tracheostomy, and decannulation in patients with Guillain-Barre-syndrome-does dysphagia matter? Muscle Nerve 59 (2) (2019) 194–200.
[4] B.R. Wakerley, N. Yuki, Pharyngeal-cervical-brachial variant of Guillain-Barre syndrome, J Neurol Neurosurg Psychiatry 85 (3) (2014) 339–344.
[5] Bath PM, Lee HS, Everton LF. Swallowing therapy for dysphagia in acute and subacute stroke. Cochrane Database Syst Rev 2018;10(10):Cd000323.
[6] S. Suntrup, T. Marian, J.B. Schroder, I. Suttrup, P. Muhle, S. Oelenberg, et al., Electrical pharyngeal stimulation for dysphagia treatment in tracheotomized stroke patients: a randomized controlled trial, Intensive Care Med 41 (9) (2015) 1629–1637.
[7] R. Dziewas, R. Stellato, I. van der Tweel, E. Walther, C.J. Werner, T. Braun, et al., Pharyngeal electrical stimulation for early decannulation in tracheotomised patients with neurogenic dysphagia after stroke (PHAST-TRAC): a prospective, single-blinded, randomised trial, Lancet Neurol 17 (10) (2018) 849–859.
[8] M. Koestenberger, S. Neuwersch, E. Hoefner, C. Breschan, H. Weissmann, H. Stettner, et al., A pilot study of pharyngeal electrical stimulation for orally intubated ICU patients with dysphagia, Neurocrit Care 32 (2) (2020) 532–538.
[9] M. Trapl, P. Enderle, M. Nowotny, Y. Teuschl, K. Matz, A. Dachenhausen, et al., Dysphagia bedside screening for acute-stroke patients: the Gugging swallowing screen, Stroke. 38 (11) (2007) 2948–2952.
[10] P. Muhle, S. Suntrup-Krueger, S. Bittner, T. Ruck, I. Claus, T. Marian, et al., Increase of substance P concentration in saliva after pharyngeal electrical stimulation in severely Dysphagic stroke patients - an Indicator of Decannulation success? Neurosignals. 25 (1) (2017) 74–87.
[11] R. Lozano, K.J. Gilmore, B.C. Thompson, E.M. Stewart, A.M. Waters, M. Romero-Ortega, et al., Electrical stimulation enhances the acetylcholine receptors available for neuromuscular junction formation, Acta Biomater 45 (2016) 328–339.
[12] M.H. Trimble, R.M. Enoka, Mechanisms underlying the training effects associated with neuromuscular electrical stimulation, Phys Ther 71 (4) (1991) 273–280 (discussion 80-2).
[13] P. Scutt, H.S. Lee, S. Hamdy, P.M. Bath, Pharyngeal electrical stimulation for treatment of Poststroke dysphagia: individual patient data meta-analysis of randomised controlled trials, Stroke Res Treat 2015 (2015) 429053.
[14] C. Fraser, M. Power, S. Hamdy, J. Rothwell, D. Hobday, I. Hollander, et al., Driving plasticity in human adult motor cortex is associated with improved motor function after brain injury, Neuron. 34 (5) (2002) 831–840.
[15] D.A. Restivo, A. Casabona, D. Centonze, R. Marchese-Ragona, D. Maimone, A. Pavone, Pharyngeal electrical stimulation for dysphagia associated with multiple sclerosis: a pilot study, Brain Stimul 6 (3) (2013) 418–423.