# Proposal for a New Exercise Method for Dysphagia with Velopharyngeal Inadequacy: A Case of Bickerstaff's Brainstem Encephalitis

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**Bickerstaff's brainstem encephalitis is an autoimmune disease with the primary lesion situated in the brainstem and three cardinal signs: ophthalmoplegia; ataxia; and impaired consciousness. A 68-year-old man was started on rehabilitation exercise 3 months after onset of Bickerstaff's brainstem encephalitis, due to remnant dysarthria and dysphagia (Functional Oral Intake Scale, level 5) after the cardinal signs of Bickerstaff's brainstem encephalitis resolved. Exercise involved using a straw in the anterior midline between the dorsal tongue and hard palate. While the patient was inhaling through the straw, the straw was blocked. After strengthening suction as much as possible, the patient was asked to immediately dry swallow at the same time that suction was stopped. Effects of exercise were examined using videofluorographic swallowing studies before and after 6 weeks of training to compare posterior and superior velar displacements and the presence of nasopharyngeal reflux. No adverse effects of exercise were encountered, and Functional Oral Intake Scale improved to level 7, with significant increases in posterior and superior velar displacement during swallowing compared with before training. In addition, nasopharyngeal reflux that had consistently been seen on swallowing before training was absent after 6 weeks of exercise. This exercise method may prove useful.**

**Key words: dysphagia, deglutition disorder, velopharyngeal inadequacy, exercise**

**List of abbreviations:**

**BBE** Bickerstaffs brainstem encephalitis
**FOIS** functional oral intake scale
**VPI** velopharyngeal inadequacy
**VFSS** videofluorographic swallowing study
**DSAS** dry swallow after suction

## INTRODUCTION

Bickerstaff's brainstem encephalitis (BBE) is an autoimmune disease with the primary lesion situated in the brainstem and three cardinal signs: ophthalmoplegia; ataxia; and impaired consciousness. In an epidemiological study in Japan, BBE was found to occur in about 100 people nationwide each year, accounting for 43% of brainstem encephalitides [1]. In addition to the three cardinal signs, patients tend to present with sensory disturbances of the peripheral limbs, diminished or absent tendon reflexes, and oropharyngeal paralysis, but very few reports have focused on the presence of dysphagia [2].

Rehabilitation was conducted for a BBE patient in whom motor disturbances in the pharyngeal stage together with velopharyngeal inadequacy (VPI) remained after immunoglobulin therapy. A novel exercise method was designed for the purpose of treating the dysphagia, based on the hypothesis that tasks facilitating velopharnygeal closure during swallowing would prove beneficial in rehabilitation for VPI. The new exercise method, in which dry swallowing is performed soon after the velum is elevated, was executed without any adverse events in this patient. This method may have beneficial actions in decreasing VPI in swallowing.

## CASE DESCRIPTION

### Subject

The patient was a 68-year-old man who developed ataxia with a chief complaint of walking difficulty after experiencing cold symptoms and was admitted to the neurology department of a university hospital. On admission, no noteworthy findings were evident on cerebrospinal MRI or cerebrospinal fluid examination. No unusual changes were seen in serum viral antibody titers. His impaired consciousness was deteriorating, and he displayed bilateral ophthalmoplegia, decreased limb tendon reflexes, and positive results for anti-GQ1b antibodies. BBE was diagnosed 3 weeks after onset, and immunoglobulin therapy was initiated. The patient did not require tracheal intubation or mechanical ventilation. While his general condition, including ataxia and ophthalmoplegia, improved steadily, dysarthria and dysphagia remained. At 3 months after onset, he was transferred to the rehabilitation department of the university hospital. At this stage, he was lucid and showed no higher brain dysfunction. In addition, no autonomic nervous system disorders including orthostatic hypotension were apparent. Hypernasality was evident and phonation time was 8 s. No motor

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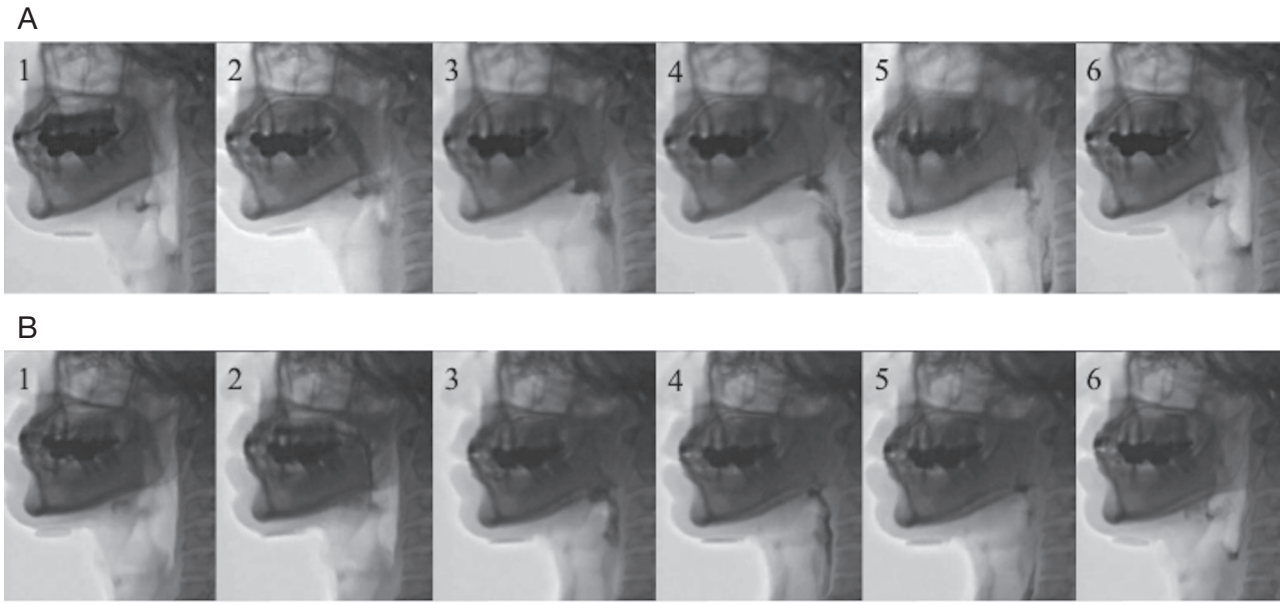


**Fig. 1** VFSS before and after DSAS
From 1 to 6, chronological changes
A. Before exercise
B. After exercise

disorder was apparent in the facial muscles or tongue. No apparent elevation of the velum was seen during phonation. His Functional Oral Intake Scale (FOIS) was level 5 (total oral diet with multiple consistencies, but requiring special preparation or compensations) [3]. Delayed initiation was not seen during pharyngeal swallowing in a videofluorographic swallowing study (VFSS), but incomplete velar elevation and decreased pharyngeal constrictor muscle function were evident. Nasopharyngeal reflux of food boluses occurred and food remained in the hypopharynx (Fig. 1A).

### Exercise Protocol for VPI: Dry Swallow after Suction (DSAS)

A straw was inserted in the anterior midline between the dorsal tongue and hard palate (straw type: diameter 4 mm × 180 mm) and the patient held it there with his mouth. While the patient was inhaling air through the straw, the tip of the straw was blocked with a finger. After strengthening suction as much as possible, the patient was asked to immediately dry swallow at the same time that suction was stopped. This was repeated eight times, representing one set. One set was performed each day, five times a week, for a total of six weeks.

The patient provided written informed consent in accordance with the Declaration of Helsinki, and all study protocols were approved by the Medical Ethics Committee of Tokai University.

### Outcome Measurements

For VFSS, the patient swallowed on command 2 ml of barium water held in the floor of the mouth. This procedure was repeated six times, with each swallow recorded on lateral-view images while the participant sat in a comfortable 60° reclining position with the neck slightly flexed. This was performed before starting and after completing exercise. Recorded frame-by-frame images (30 frames/s) of one endpoint (velar displacement) were evaluated using 2-dimensional motion analysis software (DIPP-Motion PRO 2D; DITECT Corporation, Tokyo, Japan).

### Main Outcome Measures

Posterior and superior velar displacements of the velar knee were measured relative to a Y-axis connecting the front lower corners of the second and fourth cervical vertebrae and an X-axis intersecting the Y-axis perpendicularly at the front lower corner of the fourth cervical vertebra [4] (Fig. 2).

### Secondary Outcome Measure

Lateral VFSS images were evaluated for the presence or absence of nasopharyngeal reflux.

### Data Analysis

All statistical analyses were performed using Statistical Package for the Social Sciences (SPSS) version 20.0 software (IBM, Armonk, NY). Values of $P < 0.05$ were considered to indicate statistical significance.

Wilcoxon's signed-rank test was used in comparisons of VD before and after exercise.

## RESULTS

The immediate effects of DSAS are shown in Fig. 3. The velum did not rise with air suction through the straw alone, but when the tip of the straw was blocked with a finger and suction grew stronger, the velum rose posterosuperiorly. At this time, the pharyngeal wall did not contract, but when straw suction was stopped and dry swallowing was performed before the velum dropped too much, contraction of the pharyngeal wall was obtained and velopharyngeal closure function was increased.

The patient underwent DSAS throughout the

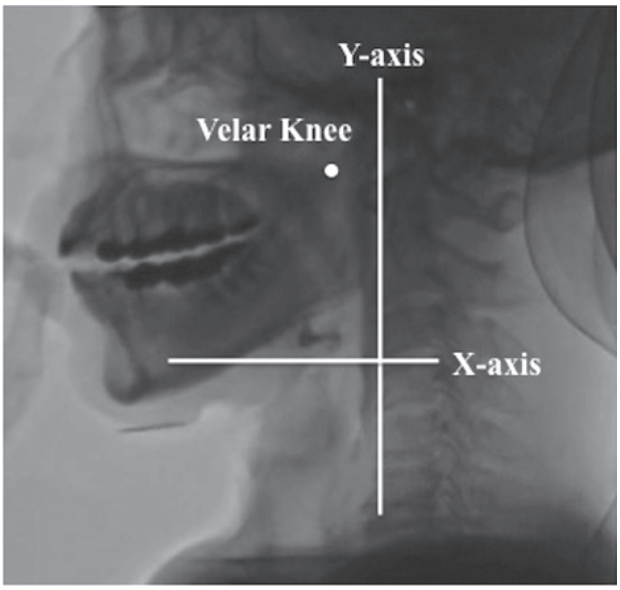


**Fig. 2** Lateral VFSS image showing X-axis, Y-axis and velar knee ( ○ ).


**Fig. 3** Immediate effect of DSAS
From 1 to 4, chronological changes
1. Air suction without blocking the straw tip
2. Strengthened suction with the straw tip blocked
3. Dry swallowing with suction stopped
4. End of dry swallowing

6-week exercise period with no adverse events, including choking during straw suction or wet hoarseness after suction, and final FOIS level improved to 7 (total oral diet with no restrictions).

**Posterior and Superior Velar Displacements**

After exercise, significant increases were seen in superior and posterior velar displacements during swallowing compared with before exercise (Table 1; Fig. 1).

**Nasopharyngeal Reflux**

Prior to exercise, nasopharyngeal reflux was seen all six times the patient swallowed (Fig. 1A-3). After completing the exercise, no nasopharyngeal reflux was seen any of the six times the patient swallowed (Fig. 1B-3).

## DISCUSSION AND CONCLUSIONS

**BBE and Dysphagia**

In addition to acute progression of the three cardinal signs of ophthalmoplegia, ataxia, and impaired consciousness, this patient showed positive results for anti-GQ1b antibodies and was diagnosed with BBE after excluding other possibilities [1].

According to an epidemiological study in Japan, the positive rate for anti-GQ1b antibodies in BBE is 75%, and the outcome for BBE showing the three cardinal signs along with positive results for anti-GQ1b antibody is good. The findings from the present study were largely consistent with these points.

In the original series of eight patients described by Bickerstaff, six of the seven surviving patients displayed complete bulbar paralysis with concomitant dysarthria and dysphagia at some stage of the illness [5]. Despite that initial report, very few cases of BBE that include instrumental swallowing evaluations have been published [2]. Oropharyngeal dysphagia may thus represent an underreported sequela of BBE, and details of the clinical course and resulting impairments are poorly understood. Whether VPI with dysphagia is typical of BBE thus remains unclear.

In the present case, improvements in VPI during swallowing were obtained around 5 months after

**Table 1** Velar displacements before and after DSAS

| Parameter | Pre-Exercise | Post-Exercise | *p* |
|---|---|---|---|
| Velar displacement (mm) | | | |
| Superior displacement | 2.9 (1.4) | 6.0 (2.9) | 0.046* |
| Posterior displacement | 0.5 (0.5) | 9.5 (2.0) | 0.028* |

Values are mean (standard deviation). DSAS: dry swallow after suction. *$P < 0.05$

onset. This supports the possibility that DSAS acts to improve function even if some of the improvement was attributable to spontaneous recovery.

**Exercise Method for VPI in Swallowing**

Blowing exercises have previously been applied with the aim of treating hypernasality accompanying VPI during phonation, but non-speech oral motor exercises proved ineffective [6]. The effects of non-swallow oral motor exercises including blowing exercises for nasopharyngeal reflux accompanying VPI in swallowing also need to be investigated, including functional improvement from generalization.

A novel exercise method was designed based on the hypothesis that tasks facilitating velopharyngeal closure would be effective in improving the nasopharyngeal reflux that accompanies VPI in swallowing. The framework for the therapeutic method emphasizes dry swallowing while improving velopharyngeal closure. The concept was to increase velopharyngeal closure by means of negative pressure load in the oral cavity from air suction using a straw. In fact, this method produced an immediate effect in posterosuperior displacement of the velum with the patient in a sitting position. However, the velum dropped when straw suction stopped, so a dry swallow needed to be elicited quickly to improve velopharyngeal closure. Moreover, no immediate effect could be confirmed in pharyngeal contraction, including Passavant's ridge, using this method alone.

Considering the above, indications for this method may be limited to VPI from disorders of velum elevation during swallowing. Obviously, this method is unsuitable for patients who do not consent to or cooperate with this exercise method and is probably unsuitable for patients in whom the elicitation of pharyngeal swallowing is delayed. Additional examinations including the tongue-holding maneuver [7] should be applied for VIP that consists mainly of decreased pharyngeal contraction.

At the same time, while this report describes the case of a single patient with mild dysphagia, he was able to continue DSAS for 6 weeks without experiencing any adverse events. The result was improvement in the VPI accompanying nasopharyngeal reflux.

To clarify the efficacy of and mechanisms underlying DSAS with greater certainty, research should be continued with designs that take into consideration the influences of gravity, including changes in posture, additional assessment with VFSS, and comparative investigations with blowing exercises.

Finally, in the future study, it is plausible to assume that DSAS is useful in the VPI improvement of convalescent stroke patients without delayed pharyngeal swallow.

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## APPENDIX : FOIS ITEMS

Level 1: Nothing by mouth.

Level 2: Tube dependent with minimal attempts of food or liquid.

Level 3: Tube dependent with consistent oral intake of food or liquid.

Level 4: Total oral diet of a single consistency.

Level 5: Total oral diet with multiple consistencies, but requiring special preparation or compensations.

Level 6: Total oral diet with multiple consistencies without special preparation, but with specific food limitations.

Level 7: Total oral diet with no restrictions.

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