

**Evaluation of Swallowing Pressure in a Patient with Amyotrophic Lateral Sclerosis
before and after Cricopharyngeal Myotomy
using High Resolution Manometry System**

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Introduction

Amyotrophic lateral sclerosis (ALS) is an extremely distressing disease, and the dysphagia often observed in the bulbar and pseudobulbar ALS is perhaps the most distressing symptom of all (1). Treatment of such dysphagia has included cricopharyngeal myotomy (2-10), but the rationale of this procedure on such patients has never been demonstrated quantitatively by novel manometric techniques.

The foundations of high-resolution manometry (HRM) were laid in the early 1990s by Clouse and Staiano (11). The advent of 'true HRM' came with the development of micro-manometric water-perfused assemblies with 36 sensors in 2006 (12). With the advancement in computer technology, esophageal HRM has been used from research to clinical practice in real time not only as conventional 'line plots', but also as 'spatiotemporal plots' (sometimes referred to as 'contour' or 'topographic' plots) displaying the direction and force of esophageal pressure activity (13).

This is the first report demonstrating the difference in swallowing pressures along the velopharynx and upper esophagus before and after cricopharyngeal myotomy in an ALS patient using this HRM system.

Case Report

A 60-year-old man visited our outpatient clinic with two years' history of progressive dysphagia. He was already diagnosed as having ALS by a neurologist in our hospital one-and-one-half years ago. As oral food intake became intolerable due to progressive aspiration, percutaneous endoscopic gastrostomy was undertaken one month ago. However, he also complained of pooling of saliva in his pharynx, and he desired to improve his complaint by surgery. On the physical examinations, fasciculation of the tongue and poor palatal movement on the right side (velopharyngeal insufficiency) were revealed. Nasopharyngeal endoscopy also demonstrated insufficient elevation of the soft palate and inadequate closure of the nasopharynx on the right side when swallowing. Videofluoroscopy (C-vision safire, Shimadzu Corporation, Kyoto Japan) with 5 ml barium liquid (100% w/v, BALITOP 100, KAIGEN Limited, Osaka, Japan) showed the pooling of barium liquid in the vallecula and the pyriform sinus and the flowing of it into the trachea (Figure 1).

After obtaining oral informed consent, we performed bilateral cricopharyngeal myotomy under general anesthesia. As his complaint has reduced one year after surgery, his quality of life has improved. On videofluoroscopy with 5 ml

barium liquid, pharyngeal transit time and cricopharyngeal opening duration (14) decreased after cricopharyngeal myotomy from 2.02 to 0.79 seconds, and from 0.84 to 0.16 second, respectively. We also measured the swallowing pressure along the velopharynx and upper esophagus using this HRM one month before and three months after the surgery.

Measurement Using High Resolution Manometry

The protocol using the HRM system (ManoScan, Sierra Scientific Instruments Inc., Los Angeles, CA) was described in our previous report in detail (15). In brief, after a local anesthesia in the nasal cavity, the catheter was inserted and fixed by taping at the nostril with the patient in a natural supine position. The patient was asked to swallow (dry swallowing) three times, and the mean values were adopted. Manometric data were analyzed using ManoView™ analysis software (Sierra Scientific Instruments Inc., Los Angeles, CA,).

Results

Figures 2 and 3 show the color graphic swallowing patterns of the

present patient before and after the surgery respectively. The color in the upper esophageal sphincter (UES) zone changed from yellow before surgery (Figure 2) to light green after surgery (Figure 3), indicating that the pressure in the UES zone decreased and the duration of pressure increase became short after surgery.

The table shows the maximum values of the resting UES pressure, the dry swallowing pressure in the velopharyngeal muscle zone and UES zone before and after the surgery. We previously reported above-mentioned values of pressure in normal Japanese men as 70.2 ± 30.0 mm Hg, (mean \pm standard deviation), 141.1 ± 73.5 and 172.7 ± 73.8 , respectively (15). Therefore, the maximum value of dry swallowing pressures at the velopharynx were poorer than normal in the present patient, and the maximum values of both the resting and dry swallowing pressures at the UES obviously decreased after cricopharyngeal myotomy from 89 to 21 mmHg, and from 171 to 75 mmHg, respectively.

Discussion

Characteristics of pharyngeal swallowing have been moderately difficult to study using conventional manometry. It is because the movement of the

soft palate and the elevation of the larynx during swallowing causes a spike-like movement along the velopharynx and upper esophagus, and also because it is extremely difficult to detect the exact pressure of a specific point by analyzing the values with only a few sensors that are widely spaced about 2 cm or more apart. We already reported the feasibility of the novel HRM system for evaluating normal swallowing function and mechanism by obtaining data of swallowing along the velopharynx and upper esophagus of normal Japanese adults, and demonstrated that this HRM can overcome these disadvantages of conventional manometric methods (15).

There have been several reports challenging evaluation of the pharyngeal pressure of the UES in ALS patients by conventional manometry (6, 16). However, there has never been a report using HRM. In the present case, with its color graphic representations, we could clearly observe the change of the swallowing patterns before and after operation along the velopharynx and upper esophagus by using HRM.

By using ManoView™ analysis software, we have also obtained the data of low swallowing pressure at the velopharynx before the surgery, which could be evidence of a cause of aspiration, and lowering the swallowing pressure at UES after

surgery, which could be evidence of the improvement in the swallowing function in the present case. Furthermore, we speculate that the relative decrease of the swallowing pressure at UES due to the surgery to the level similar to the swallowing pressure at the velopharynx might have made swallowing easy in the present case, because his swallowing pressure at UES was within normal range even before surgery. To our knowledge, this is the first report showing the effect of cricopharyngeal myotomy by demonstrating the difference in the swallowing pressure along the velopharynx and upper esophagus from the time before and after the surgery in an ALS patient using this HRM system.

The treatment of dysphagia in an ALS patient with progressive bulbar dysphagia, cricopharyngeal myotomy was first described in British literature by Mills (2). Since then, several reports have advocated the use of this procedure in patients with bulbar ALS (3-8). In the present case, the fasciculation of the tongue and the velopharyngeal insufficiency were revealed on physical examinations and nasopharyngeal endoscopy. Also, the pooling barium liquid in the vallecula and pyriform sinus and the flowing of it into the trachea were demonstrated on

videofluoroscopy. Therefore, we indicated the present ALS patient for cricopharyngeal myotomy. However, the success rate of this procedure to relieve dysphagia in ALS is 30% to 50%. Hence, there has been a diversity of opinion concerning the role of this procedure for ALS patients (17). Cricopharyngeal myotomy has been considered to be performed on the presumption that patients with bulbar ALS have spasm of the UES. This presumption is based on the work of Kirchner (18) who demonstrated that bilateral vagal nerve section at the base of the skull in the dog abolished the relaxation phase of the UES during swallowing and produced severe dysphagia during the dog's remaining life. However, the spasm of the UES was not observed in the present case. To obtain further data of swallowing along the velopharynx and upper esophagus of ALS patients using this novel HRM, we might be able to obtain a clue to prove whether the presumption is true or not.

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Figure legend

Figure 1 Videofluoroscopy showed the pooling of barium liquid in the vallecula and the pyriform sinus and the flowing of it into the trachea (Arrows) .

Figure 2. High-resolution manometry depicts swallowing pressure activity from the velopharynx to the upper esophagus in the spatiotemporal plot of the examination before operation in the present case. Time is on the x-axis (White bar indicates two seconds) and distance from nasal nostril is on the y-axis. Each pressure is assigned a color (legend right). When swallowing, we could observe constriction of the velopharyngeal muscle zone, meso-hypopharyngeal muscle zone, and the upper esophageal sphincter (UES) muscle zone. The synchronous relaxation of the UES also is obvious (A).

Figure 3. High-resolution manometry depicts swallowing pressure activity of the examination after operation in the present case. Apparent decrease in the pressure was observed in the UES zone.

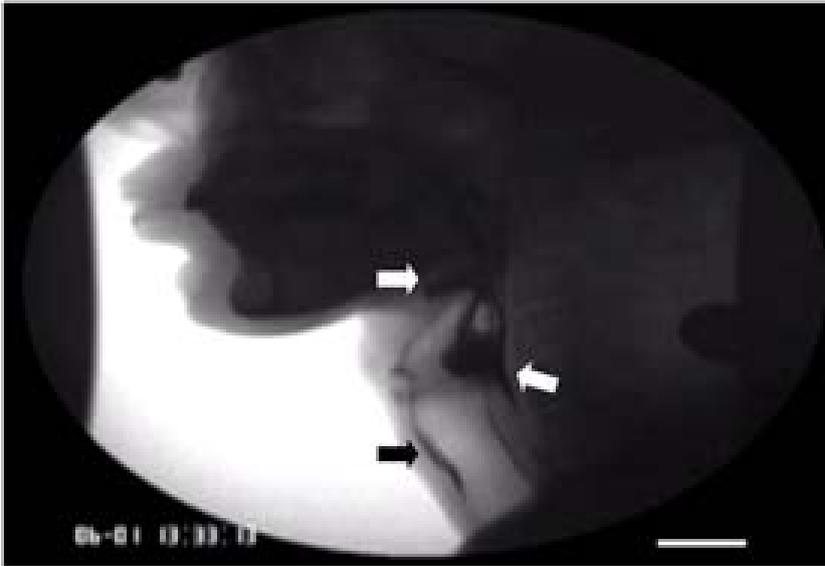


Figure 1

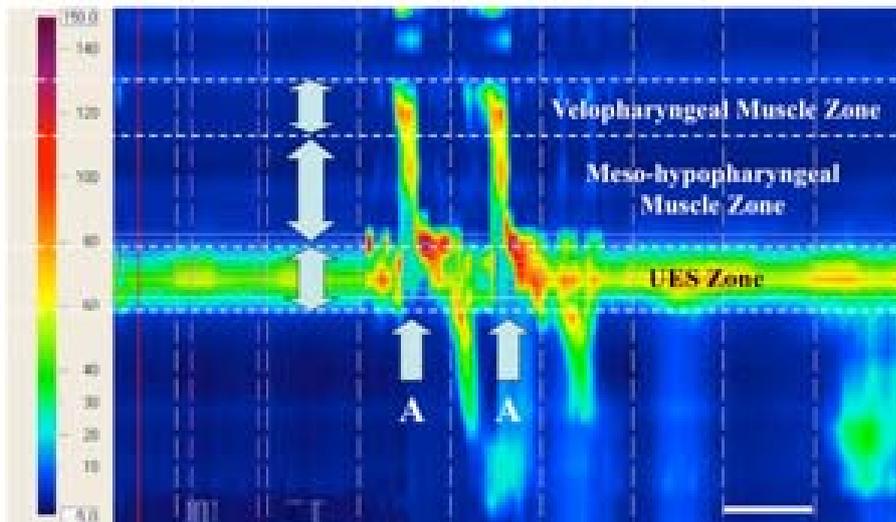


Figure 2

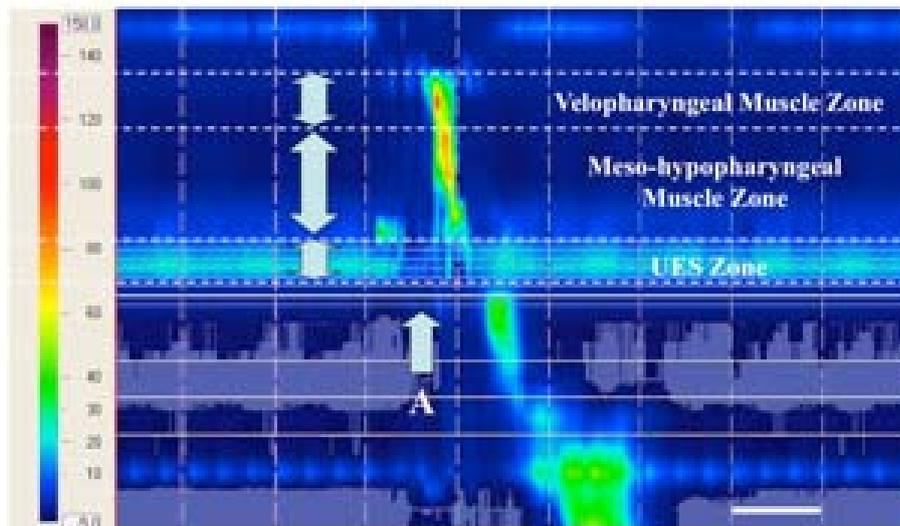


Figure 3

Table: The Maximum Values of the Resting UES pressure, Dry Swallowing Pressures in the Velopharyngeal Muscle and UES Zone at Pre- and Post-cricopharyngeal Myotomy.

	Before surgery (mmHg)	After Surgery (mmHg)
Resting UES Pressure	89	21
Dry Swallowing Pressure in the Velopharyngeal Muscle Zone	95	96
Dry Swallowing Pressure in the UES Zone	171	75

UES: upper esophageal sphincter.