



## Nasometric Scores in spinal and bulbar muscular atrophy - Effects of palatal lift prosthesis on dysarthria and dysphagia

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### ABSTRACT

Spinal and bulbar muscular atrophy (SBMA) is a hereditary neuromuscular disease affecting only males characterized by progressive muscular atrophy and weakness in bulbar and limb muscles. The present study aimed to evaluate the features of velopharyngeal dysfunction (VPD) in SBMA subjects by an acoustic analysis of speech. Twenty-three genetically confirmed patients with SBMA were enrolled and assessed their speech by measuring the nasalance score with a Nasometer II. The nasalance scores of the SBMA group was higher than that of healthy controls ( $p = .035$ ) and significantly correlated with the total score of the revised amyotrophic lateral sclerosis functional rating scale ( $r_s = -0.520$ ,  $p = .011$ ). On the basis of the results of the VPD study, the efficacy of a palatal lift prosthesis (PLP) was assessed in two patients with SBMA to treat their VPD. The PLP improved dysarthria in both cases, although the impact of the prosthesis on dysphagia was not consistent. The present study suggested that the nasalance score is a useful quantitative measurement to evaluate VPD in patients with SBMA. A PLP may improve dysarthria in SBMA patients by reducing VPD, but the clinical application of this procedure should be considered carefully in view of its possible negative effect on dysphagia.

### 1. Introduction

Spinal and bulbar muscular atrophy (SBMA) is a neuromuscular disease affecting only males, which is caused by the CAG repeat expansion encoding a polyglutamine tract in the first exon of the androgen receptor (AR) gene [1]. It is characterized by progressive muscular atrophy and weakness in bulbar and limb muscles, leading to the development of dysphagia and dysarthria in the majority of patients with SBMA [2–6]. Patients with SBMA develop dysphagia characterized by the nasal reflux of food and liquid together with decreased swallowing pressure due to insufficient elevation of the soft palate, reduced tongue movement, and poor epiglottis inversion [7]. These factors contribute to oral and pharyngeal residue and eventual penetration [7].

Various neuromuscular diseases often result in velopharyngeal dysfunction (VPD) due to poor elevation and inadequate function of the soft palate [8]. VPD is a resonance imbalance that occurs when oral sounds are produced with high nasal resonance due to poor velopharyngeal valve closure and elevation [9]. Weak or slow soft palate movements result in dysarthria of a hypernasal nature. VPD also leads to dysfunction of swallowing in addition to speech disturbance [10], since velopharyngeal closure enables the buildup of swallowing pressure and prevents material backflow into the nasal cavity in the pharyngeal swallow [11].

Although perceptual voice assessment of patients with SBMA has been reported [12], clinical studies focusing on VPD in this disease are sparse. An objective and quantitative technique to assess VPD is

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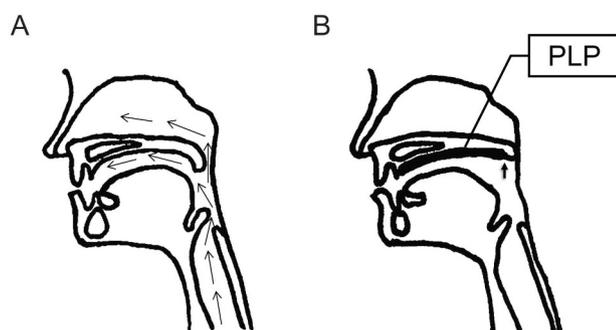
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**Fig. 1.** Velopharyngeal dysfunction (VPD) and putative effect of palatal lift prosthesis (PLP).

(A) VPD results from velopharyngeal incompetence due to poor elevation and inadequate “knee action” of the soft palate due to neurophysiological deficiency. Hypernasality as a result of resonance imbalance during speech due to poor movement of the velopharyngeal structure is characteristics of VPD. (B) The PLP is used to support to keep the soft palate normal position to improve resonance imbalance due to poor velopharyngeal valve closure and elevation.

essential for such studies, because the reliability of the perceptual assessment of hypernasality is not sufficient. Therefore, we utilized a Nasometer II, an instrument that enables the quantitative analysis of sound, to assess VPD in patients with SBMA in this study.

One of the common interventions for VPD is the application of a prosthesis [13]. A palatal lift prosthesis (PLP) is a detachable device that decreases hypernasal quality during speech and inhibits nasal regurgitation during swallowing by elevating a weakened soft palate (Fig. 1) [14–16]. This prosthetic treatment has been recommended for patients with a hypernasal quality due to pathological conditions including motor neuron diseases [14–16]. However, the efficacy of a PLP in patients with SBMA has not been reported until now. Thus, we assessed the use of a PLP in patients with SBMA and evaluated its effects on speech and swallowing dysfunction with various assessment tools, including a Nasometer II.

Previous studies suggest that the older adults with decreased muscle volume due to age-related loss of muscle bulk and strength (sarcopenia) have an increased pharyngeal lumen volume in comparison with young adults even in the healthy population [17,18]. Furthermore, increased pharyngeal lumen volume appears to rise up to worse pharyngeal constriction and this change possibly makes it more difficult to perform full pharyngeal constriction [19]. Incomplete pharyngeal constriction in swallowing was observed as the pharyngeal air space, which leads to poorer pharyngeal cleaning during swallowing [20]. The same change might occur in SBMA patients because muscle atrophy is a primary

clinical feature of SBMA patients [1–6].

In this study, we objectively assessed the characteristics of dysarthria in patients with SBMA and analyzed the relationship between the objective assessment score of hypernasality and disease severity (VPD study). Furthermore, we applied a PLP to two subjects with SBMA having VPD symptoms to assess the efficacy of the soft palate lifting procedure on dysarthria and dysphagia in this disease (PLP case study).

## 2. Materials and methods

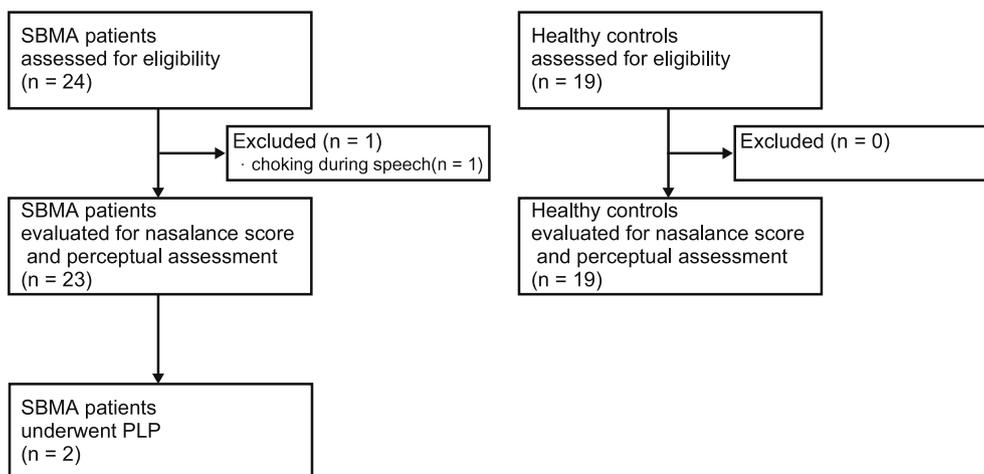
### 2.1. Subjects

A total of 23 male subjects with SBMA whose age was between 29 and 77 years were recruited consecutively to the VPD study [1]. Subjects who had tracheostomy or underwent recent intubation for any surgery were excluded. Age of onset was defined as the age at which the patient became aware of muscular weakness in any part of the body, including bulbar symptoms [12]. Disease duration from onset was also defined as the period of time between the age at onset and evaluation [12]. These SBMA patients visited the outpatient clinic of Nagoya University Hospital (Fig. 2). Nineteen age-matched healthy male Japanese speakers (37–74 years old) with normal oral structures and no history of neurological diseases were recruited as normal controls. All the data were collected at Nagoya University Hospital between August 2009 and November 2013.

In general, a PLP is considered to be effective for patients with a hypernasal quality due to insufficient elevation of the soft palate. Therefore, we selected two SBMA patients for the PLP case study based on evaluations of their swallowing and speech function. VPD, dysarthria, and dysphagia were evaluated by neurologists and Speech-Language-Hearing Therapists (SLHTs) before and after applying a PLP. SLHT is a Japanese national qualification that corresponds to the Speech Language Pathologist (SLP) in the United States. After research members concluded that the patients could markedly benefit from prosthetic treatment, patients were selected for the application of PLP (Fig. 2).

### 2.2. Acoustic analysis

A Nasometer II (model 6400; Kay Elemetrics Corp., Lincoln Park, NJ, USA), a micro-computer based instrument, was used to assess nasality. This instrument calculates the oral-nasal ratio of acoustic energy output detected by two directional microphones equipped on both sides of a sound separator board located above the upper lip. This ratio was defined as the nasalance score. In short, when the nasal resonance level increases, the nasalance score also increases. Conventionally,



**Fig. 2.** Flow chart of participants.

nasality scores have been utilized to estimate the nasality of resonance disorders resulting from cleft palate and other craniofacial disorders; however, the application range of the nasalance score has been expanded recently to neurodegenerative diseases such as Friedreich ataxia and multiple sclerosis [16,21–29]. To evaluate the nasalance score, we selected two sentences to read based on a previous study [25]: one of which was a high pressure consonant sentence (/kitsutsuki ga ki wo tsutsuku/) [26] and the other was a low pressure consonant sentence (/joui wa ooi/) [27]. These two sentences were developed by modifying the original English version [28,29] according to the characteristics of Japanese phonology [26,27]. We instructed each subject to read a pair of these two sentences three times and the average nasalance scores were recorded.

### 2.3. Auditory perceptual speech assessments

Perceptual assessments were performed to assess hypernasality, overall speech intelligibility, and audible nasal emissions according to previous reports [23,30,31]. Hypernasality was scored using a four-point scale: 1, absence of hypernasality; 2, mild degree of hypernasality; 3, moderate degree of hypernasality; and 4, severe degree of hypernasality [23]. Overall speech intelligibility was scored using a five-point scale: 1, completely unintelligible (or nearly so); 2, intelligible less than half of the time; 3, intelligible approximately half of the time; 4, intelligible more than half of the time; and 5, completely intelligible [31]. Audible nasal emission was classified as follows: +, present and –, absent [30]. Four-type speech samples were used for the perceptual assessments: the prolonged 5 Japanese vowels for assessment of speech characteristics related to VPD in vowels; reading the passage “The North Wind and the Sun” for assessment of speech characteristics related to VPD in sentence consisted of various pronunciations; oral diadochokinesia such as /papapa.../, /tatata.../, and /kakaka.../ for assessment of speech characteristics related to VPD in consonants; and light conversation in the consulting room for assessment of speech characteristics related to VPD in daily life conversation.

Recordings of the four-type samples were performed for perceptual assessment by SLHTs at a sampling frequency of 44,100 Hz and a resolution of 16 bits using a linear PCM recorder (R-09HR; Roland Corporation, Shizuoka, Japan) and a condenser microphone (ECM-23F5; Sony, Tokyo, Japan) in a silent room with a noise level below 35 dB. The distance from the microphone to the mouth was set at 15 cm [12]. The noise of room was measured by digital sound noise meter (SD-2200; FUSO, Tokyo, Japan). The sound pickup microphone of the digital sound noise meter was set the same place as the above mentioned condenser microphone. The subjects were instructed to perform the four-type speech samples at a habitual loudness and pitch.

Each speech sample was assessed independently by two SLHTs (S.T. and H.Y.) for all of the three dimensions above. The raters had an unlimited time to rate the speech samples. With regard to hypernasality and overall speech intelligibility, the raters reevaluated them if the score was different by > 1 point between the raters [23], and the average reevaluated score of the two raters was adopted. As for the assessment of nasal emission, the raters discussed their findings when their scores were different until an agreement was obtained.

### 2.4. Fabrication and application of palatal lift prosthesis

The individual anatomical form of maxilla was impressed using irreversible hydrocolloid material with custom tray to mock up a plaster stone model. The prosthesis was built up on the model using self curing acrylic resin material. After the curing process, the PLP was adjusted to individual palatal morphology by adding extra acrylic resin for extending to the soft palate site. Designs of the palatal lift for this neuromuscular diseases vary from individual to individual based on the level of activity of the palatopharyngeal musculatures. Besides, prosthetic treatment for progressive disease of patients generally must be

adjusted for disease severity. Thus modifications to the prosthesis were made on a regular basis to accommodate for the progression of the disease [14].

The adequacy of the PLP and its effectiveness in improving imbalance of resonance were confirmed clinically by assuring adequate nasal breathing and lack of dyspnea. Lateral cephalometric radiographs were taken of the patients prior and post PLP insertion to assess the adequacy of palatal lifting. The size of the gap between the superior surface of the PLP and the posterior wall of the pharynx was adjusted in consideration of speech and breathing function [32].

### 2.5. Videofluorographic swallowing study (VFSS)

Before and after applying a PLP, swallowing function was evaluated using a VFSS according to a standardized method [10,33,34]. In this examination, the patients were instructed to swallow 3 mL and 10 mL of 40% weight per volume barium sulfate three times and once, respectively [7]. Oral and pharyngeal residue, risk of aspiration, and nasal regurgitation were assessed by two independent investigators (S.T. and H.Y.). Oral and pharyngeal barium residues after initial swallowing were measured using a semi-quantitative scale. The raters reevaluated their findings if oral and pharyngeal residues were different by > 10% between the raters, and the average of the reevaluated scores of the two raters was adopted. Aspiration risk was estimated using the 8-point penetration-aspiration scale (PAS) [35]. With regard to nasal regurgitation and the PAS, any discrepancies between the investigators were discussed until an agreement was obtained [7].

We measured pharyngeal space using VFSS to confirm a relationship between oropharyngeal area [36] and swallowing function before application the PLP by using ImageJ 1.51j8 (<https://imagej.nih.gov/ij/>) [37]. Each subject was instructed to be in an upright position with their teeth in centric occlusion, lips relaxed, and head in a natural position [38]. Subject-machine distance was kept constant for each sagittal section image and a scale bar was used to calculate the area of the oropharynx [36]. The area of interest was outlined by a horizontal line from the posterior nasal spine, posterior pharyngeal wall, horizontal level of the epiglottis tip, and posterior surface of the soft palate and tongue [36].

### 2.6. Comprehensive disease severity scales

Disease severity was assessed using the revised amyotrophic lateral sclerosis functional rating scale (ALSF-RS-R) [39,40]. In addition to the total score, we analyzed the sum of three bulbar-related items (speech, salivation, and swallowing) as the bulbar subscore. We used this subscore to assess the severity of bulbar dysfunction because there was no disease-specific rating scale for SBMA at the time of the initiation of the present study [41]. We also analyzed the speech, saliva, and swallowing items separately, and defined overt subjective impairment as a score of < 4 (normal score).

### 2.7. Swallowing function questionnaires

The SWAL-QOL is a 44-item questionnaire to estimate the impact of dysphagia on quality of life (QOL) [42]. This scale was translated into Japanese and validated [43]. In the present study, section 3, which consists of 14 items (QOL in swallowing disorders), was used to assess subjective dysphagia [42]. The best and worst possible scores are 70 and 0, respectively. We defined overt subjective nasal regurgitation as a score of < 5 (never) on the 13th question of section 3 (Food or liquid coming out of your nose).

### 2.8. Genetic analysis

Genetic analysis was performed to confirm the diagnosis of SBMA. Genomic DNA was extracted from the peripheral blood of the patients

with SBMA using conventional techniques. PCR amplification of the CAG repeat in the AR gene was performed using a fluorescein-labeled forward primer (5'-TCCAGAATCTGTCCAGAAATGC-3') and a non-labeled reverse primer (5'-TGGCCTCGCTCAGGATGTCTTTAAG-3'). The detailed PCR method has been described previously [44]. Aliquots of the PCR products were combined with a loading dye and separated by electrophoresis using an autoread sequencer (SQ-5500; Hitachi Electronics Engineering, Tokyo, Japan). The CAG repeat size of the PCR standards was determined by direct sequencing, as described previously [44].

## 2.9. Statistical analyses

We analyzed the data using the non-parametric Mann-Whitney test for statistical comparisons between groups. Correlations among the parameters were analyzed using Spearman's rank correlation coefficient. To investigate the reliability of the nasalance score, the intra-class correlation coefficient (ICC) was calculated among the 3 times of reading the speech task. A receiver operating characteristics (ROC) curve was used to determine the validity of the nasalance score and hypernasality with perceptual assessment. The area under the curve (AUC), a measure of the accuracy of assessment calculated from the ROC curve, was applied for the detection of dysarthria which was defined as a score of < 4 in speech domain of ALSFRS-R. The optimal cut-off point was determined on the point where the score that added sensitivity and specificity becomes maximum [45]. *P*-values < .05 were considered to be statistically significant. Calculations were performed using the statistical software package IBM SPSS ver.25.

## 2.10. Ethics

This study was conducted in accordance with the Declaration of Helsinki, the Ethics Guidelines for Human Genome/Gene Analysis Research, and the Ethical Guidelines for Medical and Health Research Involving Human Subjects endorsed by the Japanese government. The protocol of this study was approved by the ethics committees of Nagoya University Graduate School of Medicine and Aichi Gakuin University School of Health Science. All participants gave their written informed consent prior to the interviews and examinations.

## 3. Results

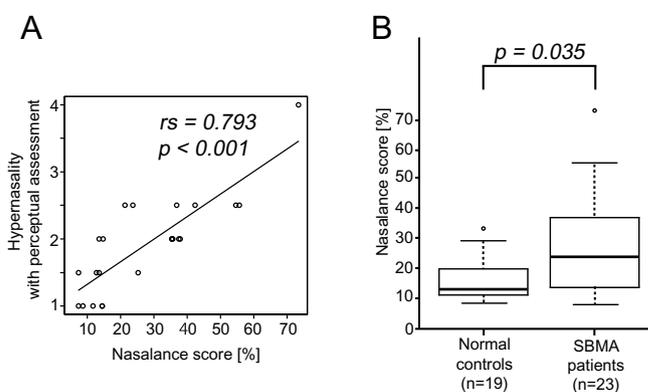
### 3.1. Baseline characteristics

The baseline characteristics of the SBMA patients are shown in Table 1. The age of normal controls (mean age = 56.9 years, range = 37–74) was not significantly different from that of the SBMA patients at evaluation (*p* = .318). The baseline characteristics of the patients were similar to those of previous reports [5–7,12,41,44].

**Table 1**  
Profiles of the patients with SBMA.

Demographic	Mean ± SD (range)
Age at evaluation, years	53.1 ± 13.4 (29–77)
Age at onset, years	40.8 ± 13.2 (14–66)
Disease duration from onset, years	12.3 ± 11.2 (2–59)
CAG-repeat length of AR gene, number	48.8 ± 2.9 (43–54)
ALSFRS-R bulbar subscores	10.2 ± 1.8 (5–12)
ALSFRS-R total score	41.0 ± 4.9 (26–48)

AR: androgen receptor; SD: standard deviation; ALSFRS-R: revised amyotrophic lateral sclerosis functional rating scale.



**Fig. 3.** Result of nasalance score.

(A) Correlation of nasalance score and hypernasality with perceptual assessment. (B) Nasalance score of patients with SBMA and normal controls. The box represents the upper and lower quartiles, and median values are represented by a thick black horizontal line within the box. The whiskers represent the maximum and minimum values, which include the width between the first quartile  $-1.5 \times$  interquartile range and third quartile  $+1.5 \times$  interquartile range. Outlier are shown as a small circles.

### 3.2. Auditory perceptual voice assessments of patients with SBMA (VPD study)

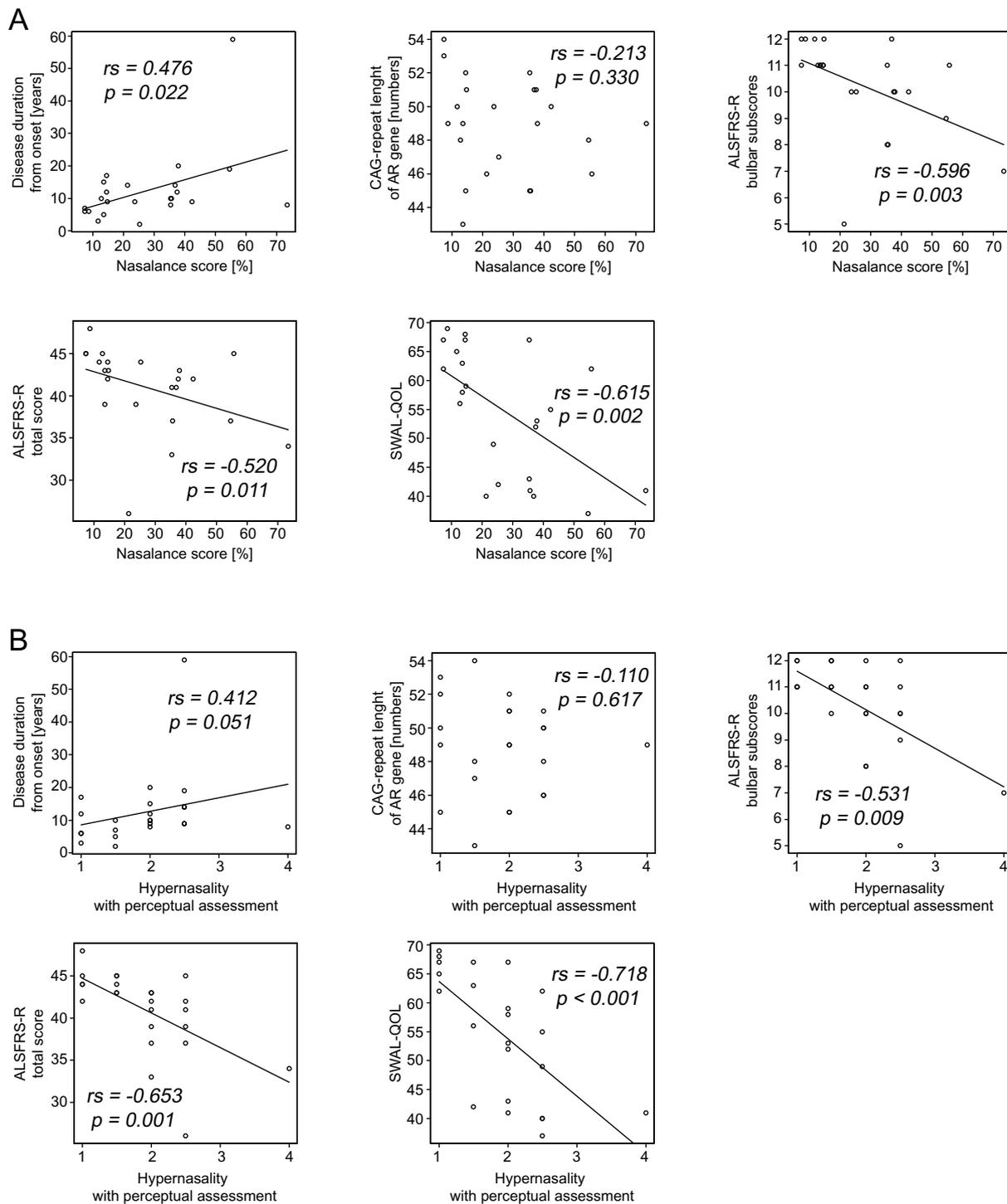
As a result of the perceptual assessment of speech by two SLHTs, the mean score of “hypernasality” was 1.91 (standard deviation [SD] = 0.72, range = 1.0–4.0), being 2.0 (mild hypernasality) or higher in 14 out of 23 subjects with SBMA. The mean score of “Reduced overall speech intelligibility” was 4.33 (SD = 0.75 and range = 2.0–5.0), being 4.0 (intelligible more than half of the time) or lower in 9 out of 23 subjects with SBMA. “Audible nasal emission” was detected in 43.5% of patients.

### 3.3. Nasalance score in patients with SBMA (VPD study)

The nasalance score correlated well with hypernasality measured with perceptual assessment (Fig. 3A). The nasalance score of the patients with SBMA was significantly higher than that of the normal controls (Fig. 3B). The nasalance score showed a strong intra-class correlation among 3 times of reading speech task (ICC = 0.979, 95%CI = 0.966–0.988, *p* < .001).

### 3.4. Relationship between hypernasality and bulbar function in patients with SBMA (VPD study)

The degree of hypernasality measured with both methods, Nasometer and perceptual assessment, was correlated with clinical background such as ALSFRS-R bulbar-related subscores and swallowing scale (SWAL-QOL) (Fig. 4). As for subjective impairments assessed by each bulbar item of the ALSFRS-R, the nasalance score of the patients with subjective dysarthria was significantly higher than that of those without subjective dysarthria (Fig. 5A). Conversely, there was no difference in the degree of perceptually assessed hypernasality between the patients with and without subjective dysarthria (Fig. 5B). Both subjective salivation and dysphagia tended to be associated with a higher nasalance score, although the difference was not statistically significant (Fig. 5A). Both the nasalance score and degree of perceptual assessment, which were used to assess hypernasality, were higher in patients with subjective nasal regurgitation than in those without (Fig. 5C).



**Fig. 4.** Relationship between nasal quality and clinical background. (A) Relationship between nasalance score and clinical background. (B) Relationship between hypernasality with perceptual assessment and clinical background. rs, Spearman's rank correlation coefficient; AR, androgen receptor; ALSFRS-R, revised amyotrophic lateral sclerosis functional rating scale; SWAL-QOL, swallowing-quality of life questionnaire.

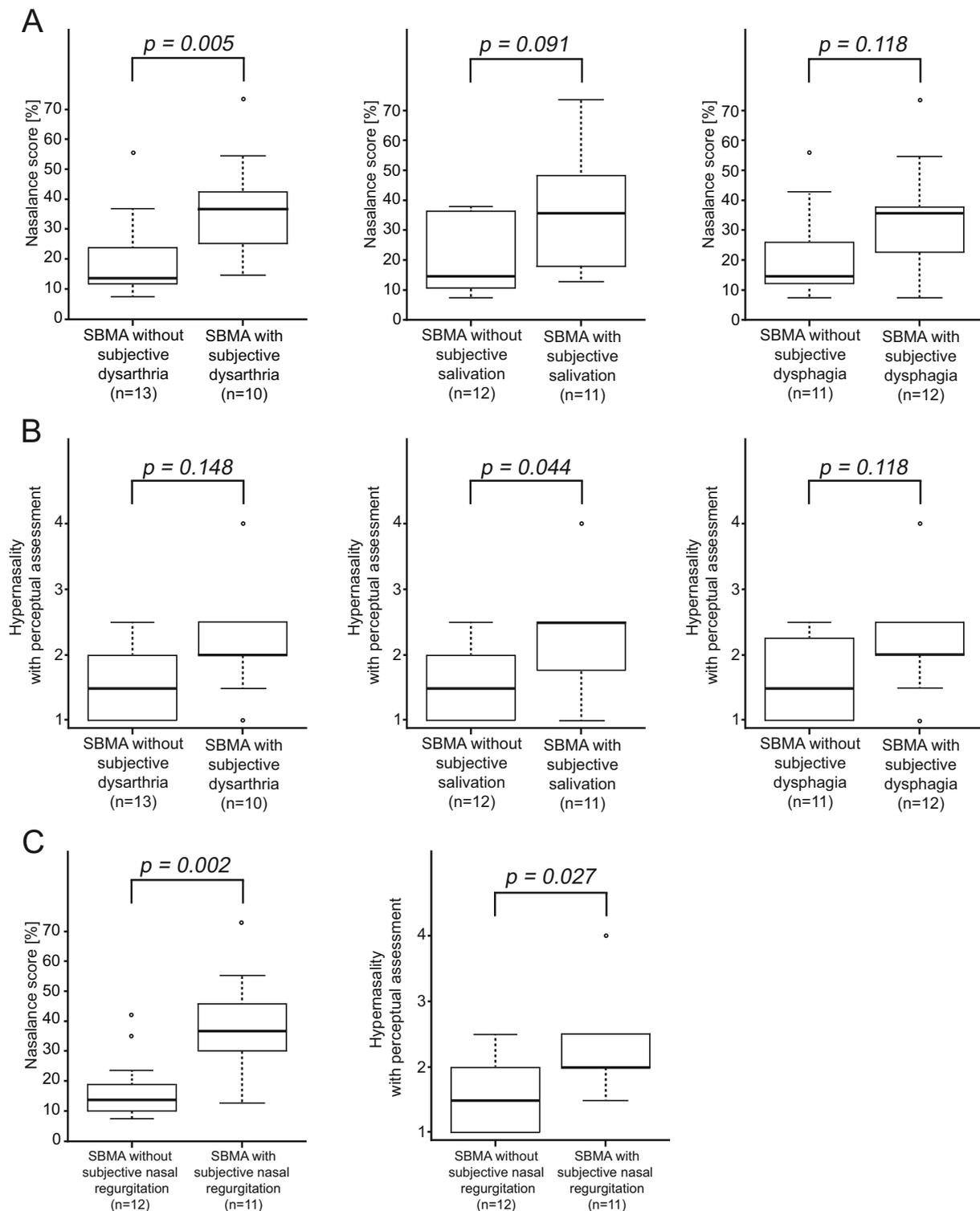
**3.5. Validity of nasalance score and hypernasality with perceptual assessment (VPD study)**

ROC curve analysis was applied between the SBMA patients with and without subjective dysarthria (Fig. 6). The results demonstrated that nasalance score was significantly higher in the patients with subjective dysarthria than in those without (AUC = 0.838, AUC 95% IC = 0.672–1.000, optimal cut-off point = 24.420%, sensitivity

= 0.800, specificity = 0.769,  $p = .006$ ). However, this finding was not observed as for hypernasality with perceptual assessment (AUC = 0.685, AUC 95% IC = 0.463–0.906, optimal cut-off point = 1.750, sensitivity = 0.800, specificity = 0.538,  $p = .137$ ).

**3.6. Case reports of SBMA patients with a PLP (PLP case study)**

Two patients with SBMA who had moderate to severe VPD were



**Fig. 5.** Comparison of nasal quality between subjects with and without subjective bulbar symptoms measured with the ALSFRS-R.

(A) Nasalance score. (B) Hypernasality judged by perceptual assessment. (C) Comparison of nasal quality between the SBMA subjects with and without subjective nasal regurgitation. Median values are represented by a thick black horizontal line within the box, and the box represents the upper and lower quartiles. The whiskers represent the maximum and minimum values, which include the width between the first quartile  $-1.5 \times$  interquartile range and third quartile  $+1.5 \times$  interquartile range. Outliers are shown as small circles.

included in the PLP case study. Dysarthria and dysphagia due to VPD were evaluated by two SLHTs before and after applying a PLP. The evaluation was performed on the subjects before applying a PLP (pre-PLP) and after applying a PLP in condition with PLP (post-PLP) (Table 2). In the pre-PLP condition, these two cases had some different

characteristics. For instance, *Case 2* had a larger oropharyngeal space (oropharyngeal area) than *Case 1*.

**Case 1.** A 39-year-old male subject with SBMA with hypernasality and nasal emissions during speech and awareness of food and liquid flowing into his nasal cavity. He noticed muscle weakness of his limbs at the age

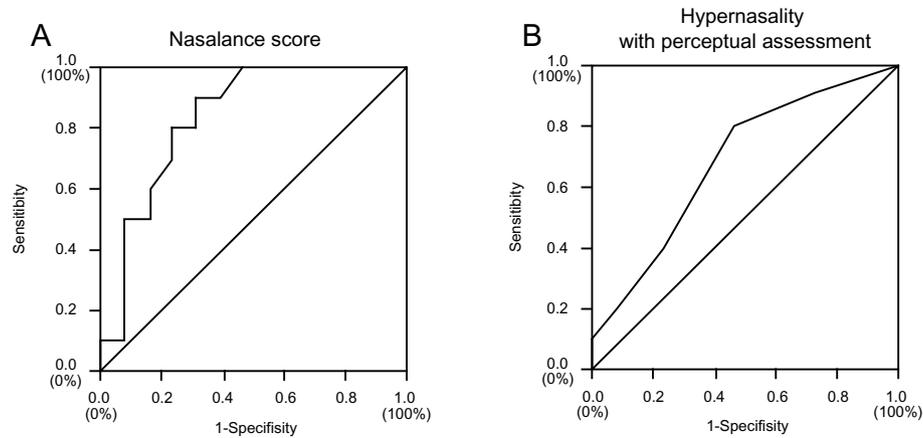


Fig. 6. Efficacy of Nasometer and perceptual assessment for describing VPD.

Receiver operating characteristics (ROC) curve analysis of SBMA patients with and without subjective dysarthria using nasalance score (A) and perceptual assessment by SLHTs (B).

Table 2

Clinical background and effects of a PLP in two patients.

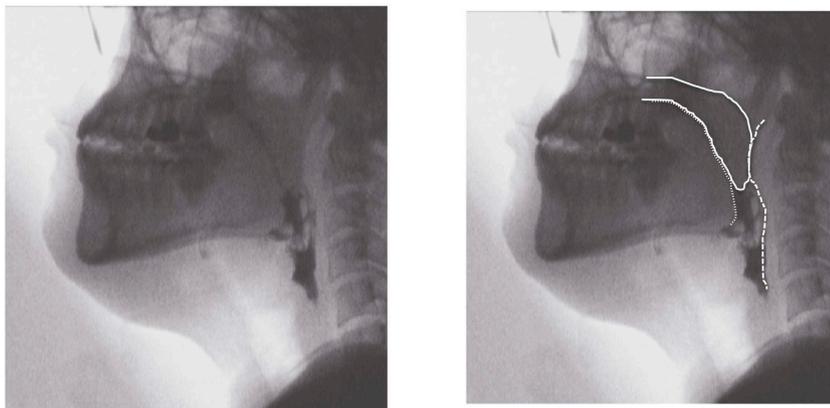
Items	Case 1		Case 2			
	Pre-PLP (Base line)	Post-PLP (1 year and 8 months later)	Pre- PLP (Base line)	Post-PLP (1 year and 4 months later)		
Age at evaluation, years	39	41	56	57		
Age at onset, years	31		48			
Disease duration from onset, years	8	10	8	9		
CAG-repeat length of AR gene, numbers	52		49			
ALSFRS-R						
Speech	3	3	2	3		
Salivation	2	3	3	2		
Swallowing	3	3	2	2		
Bulbar subscores	8	9	7	7		
Total	33	36	34	32		
Speech	Perceptual assessment by SLHTs	Intelligibility <sup>a</sup>	5.0	5.0	2.0	4.0
		Hypernasality <sup>b</sup>	2.0	1.0	4.0	3.0
		Nasal emission <sup>c</sup>	+	-	-	-
Swallowing (VFSS)	Nasalance score		% 35.3	32.2	73.3	65.5
		3 mL (average of three times)	Pharyngeal residue <sup>d</sup>	% 20.0	5.8	9.5
		Intraoral residue <sup>d</sup>	% 13.3	6.7	18.3	13.3
		Nasal penetration <sup>e</sup>	0/3	0/3	0/3	0/3
		PAS <sup>f</sup>	1.0	1.0	1.0	3.7
	10 mL	Pharyngeal residue <sup>d</sup>	% 5.0	3.5	5.0	35.0
		Intraoral residue <sup>d</sup>	% 20.0	7.5	20.0	15.0
		Nasal penetration <sup>e</sup>	0/1	0/1	0/1	0/1
		PAS <sup>f</sup>	1	1	2	8
	Oropharyngeal area	cm <sup>2</sup>	7.38	NT	10.07	NT

PLP: palatal lift prosthesis; AR: androgen receptor; ALSFRS-R: revised amyotrophic lateral sclerosis functional rating scale; SLHT: Speech-Language-Hearing Therapist; VFSS: videofluorographic swallowing study; PAS: penetration-aspiration scale; NT: not tested. Interpretation of each rating scales as follow; a. From 1 = completely unintelligible (the worst) to 5 = completely intelligible (the best); b. From 1 = absence of hypernasality (the best) to 4 = severe degree of hypernasality (the worst); c. + = presence, - = absence; d. 0 (the best), 2, 5, 10, 20, 30, 40, 50, 60, 70, 80, 90, and 100% (the worst); e. Number of finding this phenomenon during trial; f. From 1 = material dose not enter airway (the best) to 8 = material enters the airway, passes below the vocal folds, and no effect is made to eject (the worst).

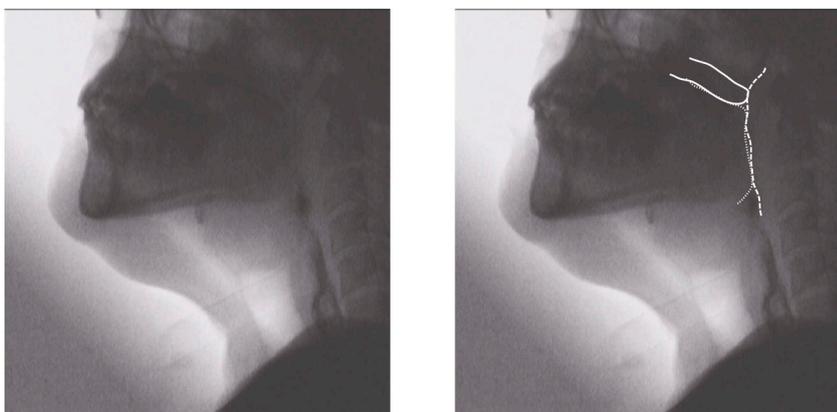
of 31 years and was diagnosed with SBMA by genetic analysis (CAG repeat length of 52 in the AR gene) (Table 2). His speech and swallowing symptoms emerged at 30 and 37 years of age, respectively. The Nasometer II and perceptual assessments by two SLHTs indicated that the PLP slightly decreased his nasalance score and improved his dysarthria. Swallowing function evaluated with VFSS showed no clear change following PLP attachment (Table 2). In pre-PLP condition, his soft palate was located between the base of the tongue and pharyngeal wall. Attaching the PLP (post-PLP) resulted in an increase of the maximum elevated level of the soft palate and more obvious contact with the base of the tongue and posterior wall of the pharynx during the swallowing reflex (Fig. 7).

**Case 2.** A 56-year-old subject with SBMA with hypernasality and distortion of explosive sounds during speech and awareness of the penetration of food or liquid into his nasal cavity. He noticed muscle weakness of his lower limbs at the age of 48 years and was diagnosed with SBMA by genetic analysis (CAG repeat length of 49 in the AR gene) (Table 2). Although the nasalance score and dysarthria was improved by the PLP, its effect on dysphagia was not consistent. Although his speech improved when the PLP was attached, both the PAS and pharyngeal residue deteriorated. With 3 mL of barium (average of 3 times), the PAS worsened from 1.0 to 3.7 and the pharyngeal residue also increased from 9.5% to 65.0%. Place of pharyngeal residue was changed from pyriform sinuses to throughout the pharynx. With 10 mL of barium, the PAS worsened from 2 to 8 and the pharyngeal residue

A



B



**Fig. 7.** Images of a VFSS of *Case 1*.

(A) In “Pre-PLP” condition, this case showed chin up and a decline of maximum elevating level of soft palate. Soft palate sat between the base of tongue and the posterior wall of pharynx during swallowing reflex. As a result, the movement range of tongue decreased. (B) When the PLP was attached to the subject, his soft palate was raised. As a result, the range of the posterior tongue movement increased to contact with pharyngeal wall to generate swallowing pressure. The white lines are the end of the hard palate and soft palate. The white broken lines are the posterior wall of the pharynx. The white dotted lines are the base of the tongue.

also increased from 5.0% to 35.0%. Although the amount of pharyngeal residue increased by using the PLP, place of pharyngeal residue was not clearly changed, that is, the pharyngeal residue occurred throughout the pharynx in both pre- and post- PLP conditions. The VFSS under the condition with the PLP showed that the PLP widened the space surrounded by the soft palate, base of the tongue, and posterior wall of the pharynx by lifting the soft palate (Fig. 8). This unnecessary space was not seen in the pre-PLP condition and it appears to underlie the deterioration of swallowing function. VFSS indicated that this pharyngeal residue might be caused by incomplete contact of the posterior tongue with the soft palate. The presence of pharyngeal residue accompanied by incomplete closure of the laryngeal vestibule during the swallowing reflex might lead to aspiration of pharyngeal residue.

#### 4. Discussion

Our VPD study demonstrated that one of the most important features of dysarthria in patients with SBMA is hypernasality due to incomplete lifting of the soft palate. Measurement of the nasalance score using a Nasometer II is a useful marker of hypernasality in patients with SBMA. Radiographic and endoscopic examinations have been used to evaluate VPD, but these methods have disadvantages such as excessive radiation exposure or the necessity for skillful technique [13].

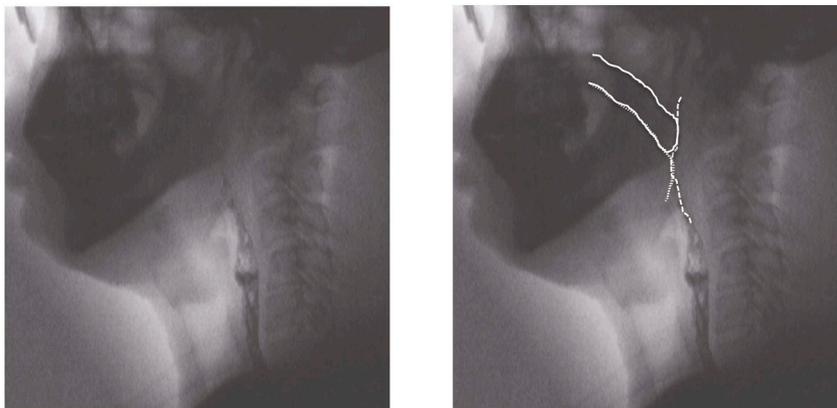
To avoid an ambiguous assessment of hypernasality, we utilized the nasalance score using a Nasometer II, which is an objective and quantitative technique to assess VPD. In the present study, the nasalance score measured with this simple noninvasive equipment correlated well

with disease duration and severity scales (ALSFERS-R and SWAL-QOL) that have been used widely to assess patients with SBMA. A significant difference in the nasalance score was also observed between patients with and without subjective dysarthria and subjective nasal regurgitation when consuming a daily meal. The VPD study also showed that the nasalance score was correlated with the severity of hypernasality as evaluated by perceptual assessment, as well as the subjective symptoms of VPD.

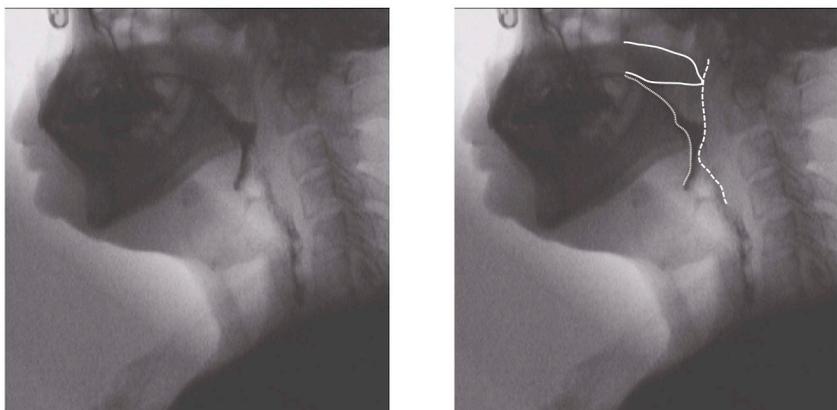
Among the available non-invasive examinations, the perceptual assessment of hypernasality is practical for the diagnosis of VPD [13]; however, it has several critical limitations including low inter-evaluator reliability [8]. In addition, perceptual assessment of subjects with both dysphonia and hypernasality is very difficult because a breathy voice tends to sound hypernasal [46]. In the present study, objective analysis using Nasometer, but not perceptual assessment, detected a significant difference in the degree of hypernasality between the SBMA patients with and without subjective dysarthria. These results are likely stem from the influences of dysphonia on the perceptual assessment of hypernasality, given that, in accordance with these results, 70% of patients with SBMA were confirmed to have dysphonia in perceptual voice analysis using the GRBAS scale [12]. It is possible that Nasometer II is more specific assessment method of hypernasality during speech than perceptual assessment. These data indicated that the nasalance score is a useful marker for the assessment of VPD in patients with SBMA.

As mentioned above, a principal feature of VPD in the patients with SBMA was hypernasality due to incomplete lifting of the soft palate, which is why we used a PLP in two patients with VPD and assessed its efficacy using various assessment tools including a Nasometer II. As a

A



B



**Fig. 8.** Images of a VFSS of *Case 2*.

(A) In “Pre-PLP” condition, this case showed a decline of maximum elevating level of soft palate. Soft palate didn't move at all during VFSS. Soft palate hung down as so to cover the contact point of the base of tongue and the posterior wall of pharynx at a swallowing reflex. (B) The PLP increased the maximum elevating level of the soft palate. In comparison with *Case 1*, the PLP produced extra space surrounded by the soft palate, base of the tongue, and posterior wall of the pharynx. The white lines are the end of the hard palate and soft palate. The white broken lines are the posterior wall of the pharynx. The white dotted lines are the base of the tongue.

result, the PLP showed different effects on dysarthria and dysphagia in both patients. The prosthesis improved hypernasal quality, but not swallowing, in both cases. Speech was improved probably because the PLP improved resonance imbalance during the production of non-nasal vowels and consonants. In contrast, the effects of the PLP on dysphagia were inconsistent. It worsened dysphagia in *Case 2*, while *Case 1* showed no detectable change of dysphagia following the application of the PLP. Normal swallowing requires sufficient swallowing pressure to send the bolus out of pharynx. Lifting the soft palate improves resonance imbalance during speech; however, it also results in the emergence of an unnecessary extra space surrounded by the soft palate, base of the tongue, and posterior wall of the pharynx because the musculature of patients with SBMA tends to be atrophied. The unnecessary space thus indicates incomplete full pharyngeal constriction, which possibly leads to increase pharyngeal residue.

Previous reports demonstrated that a PLP is effective for dysarthria due to neurological disease [14–16]. However, the effect of a PLP on dysphagia due to neurological disease has been scarcely documented, except for a few case series reports [15]. A PLP has been shown to be an effective intervention for amyotrophic lateral sclerosis patients with dysarthria and for stroke patients with both dysarthria and dysphagia [14,15], but its efficacy in other neuromuscular diseases remains elusive. The present study demonstrated that the effect of the PLP on speech was consistent, but its effect on swallowing dysfunction depended on disease stage, which might be related to the extra oropharyngeal space caused by atrophy of the bulbar muscles. Based on the result of the present study, the PLP may be used for treating speech disturbance in SBMA patients, but it should not be used in those with severe swallowing dysfunction. If using PLP, SBMA patients would be

advised to remove it during feeding times and maintain it throughout the day where speech is mandated. A limitation of the present report was that only two subjects were recruited to the PLP case study. In order to confirm the efficacy of a PLP, further studies will be needed with larger numbers of SBMA patients with VPD.

## 5. Conclusions

The present study indicated that the nasalance score is a useful marker for the assessment of symptoms related to VPD and the effect of prosthetic intervention. The PLP improved the hypernasality of patients with SBMA because it improves resonance imbalance during speech by elevating the soft palate. Nevertheless, the effect of the PLP on dysphagia was not consistent among the SBMA subjects, indicating the need for the careful consideration of PLP application in SBMA.

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## Declaration of Competing Interest

Drs. Tanaka, Hijikata, Yamada, Ito, Nakayama, Kurita, Banno, Suzuki, and Yamamoto declare that they have no conflict of interest.

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Mr. Yogo declares that he has no conflict of interest.

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