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ORIGINAL REPORT

STRATEGIES FOR LEARNING GLOSSOPHARYNGEAL BREATHING IN BOYS WITH DUCHENNE MUSCULAR DYSTROPHY: A FEASIBILITY CASE SERIES

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Objective: **To propose alternative learning strategies for glossopharyngeal breathing in patients with Duchenne muscular dystrophy (DMD) and healthy men.**
Design: **A feasibility study with small case series.**
Subjects: **Five boys with DMD and 7 male physical therapists as healthy controls who had not learned glossopharyngeal breathing.**
Methods**: Participants were instructed in a glossopharyngeal breathing protocol, including induction methods comprising sucking motions and phonation with inhalation. The protocol consisted of 1–6 sessions (10–15 min each; total 60 min). Criteria for glossopharyngeal breathing mastery were vital capacity with glossopharyngeal insufflation (VC_{GI})/VC ratio > 1.10 for the DMD group and > 1.05 for the Healthy group. Feasibility outcomes were time required for mastering glossopharyngeal breathing, self-reported outcomes, adverse events and drop-outs.**
Results: **All participants learned glossopharyngeal breathing within the allocated 60 min. Mean VC_{GI}/VC ratio was 1.31 for the DMD group and 1.09 for the Healthy group. No adverse events or drop-outs were encountered during the protocol. In most cases, self-reported outcomes showed that motivation increased and difficulty decreased over time.**
Conclusion: **Induction methods for sucking motions and phonation with inhalation for glossopharyngeal breathing learning are feasible. This paper proposes alternative strategies for glossopharyngeal breathing learning in boys with DMD and their instructors.**

Key words: neuromuscular disease; breathing exercise; insufflation; decision-making; physical therapy.

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LAY ABSTRACT

Glossopharyngeal breathing is an active breathing manoeuvre that involves the subject autonomously pistoning boluses into their lungs without the use of any device, providing a form of positive-pressure breathing for type of patients with restricted breathing. A strategy for learning glossopharyngeal breathing has not yet been established and the manoeuvre is often difficult to learn for patients with Duchenne muscular dystrophy (DMD). All subjects in this study (5 boys with DMD and 7 healthy men) were able to master glossopharyngeal breathing. Two types of guidance, regarding sucking motions and phonation with inhalation, might facilitate boys with DMD and healthy controls in learning glossopharyngeal breathing. This paper proposes a glossopharyngeal breathing strategy to help clinicians by setting out a novel protocol for learning glossopharyngeal breathing more effectively.

Duchenne muscular dystrophy (DMD) is an X-linked recessive neuromuscular disorder caused by mutations in the dystrophin gene, characterized by progressive muscle weakness and wasting, including the respiratory and cardiac muscles (1). In a recent systematic epidemiological review, DMD was reported to occur predominately in males, with a birth prevalence of 15.9–19.5 cases per 100,000 newborn males and median survival of 24–26 years (40.9 years for patients born after 1970) (2). Respiratory management in DMD can decrease respiratory complications and prolong survival (3, 4). A recent review found that a structured, anticipatory approach to respiratory management requires monitoring of respiratory muscle strength, as well as initiation of lung volume recruitment (LVR), assisted coughing, nocturnally-assisted ventilation and, eventually, daytime ventilatory support (5, 6). In an international consensus opinion, LVR was considered to limit chest wall contracture and lung restriction, increase cough peak flow and voice volume, promote lung growth and impede chest deformity among children with neuromuscular disease (NMD) (7). Continuous implementation of LVR in DMD can delay decline in vital capacity (VC) (8). LVR is applied using a bag valve mask or volume ventilator, mechanical insufflation-exsufflation device, and glossopharyngeal breathing (GPB) (5, 9, 10).

GPB (12) is an active breathing manoeuvre that involves the subject independently pistoning air boluses into their lungs without the use of any device, providing a form of positive-pressure breathing (12, 13). The technique consists of active step-by-step inhalation using the tongue, soft palate, fauces, pharynx, larynx, and vocal cords (11, 13). This manoeuvre is currently known as a specific breathing technique for individuals with NMD and spinal cord injuries and other situations, including in sports medicine (14–16). Patients who have learned GPB may achieve a maxi-

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mum insufflation capacity (MIC) comparable to that achieved with LVR using a bag valve mask (10, 17). Thus, GPB can be used by patients with decreased VC, in order to cough more effectively, increase speech volume, maintain pulmonary compliance, and prevent atelectasis (18, 19). Learning GPB is a crucial skill in individuals with respiratory muscle paralysis, and its application should be considered (20). Learning GPB has been recommended under recent guidelines for patients with NMD (21).

Learning GPB can be difficult and time-consuming (21–23). According to Bach et al. (10) the proportion of patients who succeed in mastering GPB with DMD was as low as 26.9%, compared with 94.9% for LVR. Nevertheless, the mastery rate for GPB appears markedly higher among physical therapists (24) and elite swimmers (15, 16). Certainly, it should be possible for patients with DMD patients, in whom glottal closure capability is maintained, to learn GPB (21). It may be more efficient for patients with DMD to learn GPB when respiratory or glossopharyngeal functions remain (11), as the transition to assisted ventilation is very likely.

Methods for learning GPB typically involve the following steps: description of GPB (13, 24–27); instruction in "swallowing air" (28); observation of GPB performance and actual technique on video (10, 13, 20, 25–27, 29, 30); imitation of an instructor or GPB-competent individual (10, 13, 15, 27, 30); use of visual feedback or instruction manuals (25, 26, 29), vocalization (13, 23, 25), and tutoring by the therapist (24, 30, 31). Current GPB learning methods basically centre on clinical practice using trial-and-error observation and imitation by patients. The learning methods are not uniform, due to variations in GPB techniques (32). Better learning methods should be developed in order to gain significant benefits for many patients (18, 21). In addition, therapists providing instruction to patients should also have learned GPB, but the strategies and time required to master GPB in the intact population are unclear (22, 24). Research into systematic teaching of GPB and its validity, and options for the methodology, are lacking.

A series of protocols, including 2 alternative strategies from prior reports and clinical practice, was hypothesized to be viable for mastering GPB. The protocol for learning GPB could be used for efficient, short-term mastery by patients with DMD with remaining respiratory function. The objectives of this study were therefore to propose alternative GPB learning strategies for patients with DMD and physical therapists, and to determine the feasibility of mastery of GPB.

METHODS

Design

The study was designed as a case series and a feasibility study. Session-by-session tracking of 5 case studies was employed for a DMD group using our protocol for GPB mastery. The main outcome was whether GPB was mastered. Feasibility was verified by referring to the time required, self-reported outcomes, and safety throughout the daily protocol. Physical therapists who had not previously learned GPB were included as a healthy control group, and the same protocol was performed.

Participants

Participants were recruited in the Department of Rehabilitation Medicine at National Hospital Organization Higashisaitama National Hospital between January 2018 and June 2019.

Boys with DMD were recruited as consecutive patients by a single therapist, according to the following criteria. Inclusion criteria were: age $\geq$16 years; diagnosis of DMD through DNA analysis and/or muscle biopsy, and electromyography; inpatient status or one visit per week during the study period; and ability to implement LVR by bag valve mask. Exclusion criteria were: present competence with GPB; presence of mental retardation or severe cardiac failure; or indwelling tracheostomy tube. Ventilator use and history of GPB practice were confirmed prior to protocol implementation in the DMD group.

A group of male physical therapists, other than the supervisors, was recruited from a convenience sample as healthy controls (Healthy group). The inclusion criteria were: age 20–40 years; and no prior involvement in GPB instruction. Exclusion criteria were: present competence with GPB; pain or disability in the glossopharyngeal area; or respiratory disease or impairment.

All study protocols were approved by the local research ethics committee of the National Hospital Organization Higashisaitama National Hospital and were consistent with the principles of Declaration of Helsinki. All subjects provided written informed consent to participate. This study was registered with the UMIN Clinical Trials Registry (UMIN-CTR number: UMIN000030422).

Intervention

The explanation of the mechanics underlying GPB represented original material that incorporated findings and descriptions from previous reports (Fig. S1[1]) (11, 13, 18, 21–23, 32, 33). The main mechanisms of GPB involve repetition of the following coordinated movements (32, 33): (*i*) elevating the soft palate for nasopharyngeal closure; (*ii*) securing air space by flattening of the tongue and downward movement of the larynx; (*iii*) sealing the palato-glossal cavity with the tongue and palate and delivering the air to the pharynx; and (*iv*) glottal closure after momentary glottal opening, accompanied by upward movement of the larynx. Common factors making GPB difficult to learn appear to be inadequate valve closure (20, 22) and upward and downward movements of the larynx (10, 33). The main strategy in our protocol was to guide proper positioning and actions of the glossopharynx using 2 induction methods, to identify air leaks from the nose and/or mouth and to achieve appropriate valve closure.

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Table I. Brief version of the learning protocol for glossopharyngeal breathing (GPB)

Preparation step	
Lectures	
1	Description of significance and benefit
2	Illustration of mechanism
3	Watching demonstration video provided by patients who had already mastered GPB
4	Watching real-time demonstration by the supervisor
Practice step	
Conventional strategies	
1	Imitation method
2	Simple instruction method
Alternative strategies	
3	Induction of sucking motion
4	Induction of phonation with inhalation

The protocol used in this study included 2 specific instructional strategies and comprised the following steps (Table I, see Fig. S2[1] for greater detail): (*i*) conventional methods, including observation of an individual who had mastered GPB, explanation of the mechanics underlying GPB, and use of imitation or imagination; (*ii*) induction of sucking motions (Fig. 1A); and (*iii*) induction of phonation with inhalation (Fig. 1B). Induction of sucking motion was a method using innate behaviours similar to those in GPB and imitating the sucking motions seen in breast-feeding. In the early stages of training, deflation of the small bag through a straw by sucking motions can reflect the stacking in the lungs when using a nose clip. Induction of phonation with inhalation used "voiceless pronunciation" to lead to proper tongue positioning and glossopharyngeal insufflation.

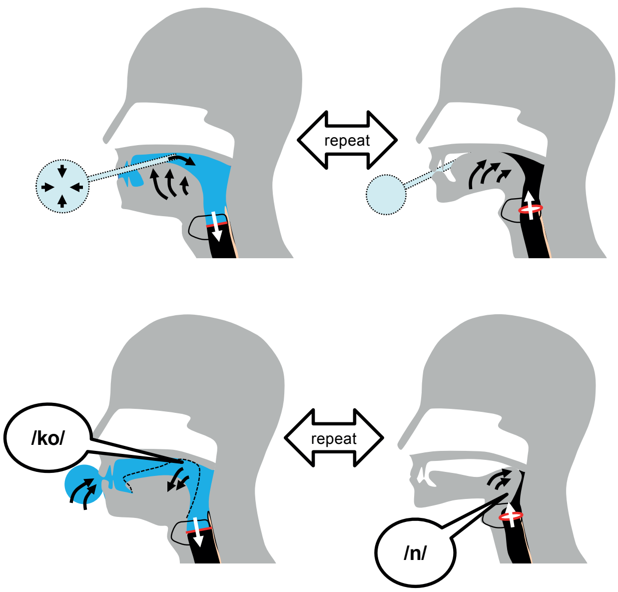

Fig. 1. Two induction methods for learning glossopharyngeal breathing (GPB). (A) Induction of sucking motion. *Left:* push the sucked air backwards while squashing the tongue against the hard palate (at this time, the air bag should deflate). *Right:* Gulp and keep the sucked air within the laryngopharyngeal cavity (at this time, the air bag should not re-inflate if no air leaks are present). (B) Induction of phonation with inhalation. *Left:* Phonation with inhalation: make the shape required for voiceless pronunciations of "/k/" and "/o/" (other phonemes are /ku/, /ka/, /to/, /tu/, /ta/). *Right:* Swallowing while breath-holding: trap the air with glottic closure and laryngeal elevation, while making the shapes for voiceless pronunciation of "/n/" or "/m/".

Glossopharyngeal movement in the first half of insufflation is ideally represented by uttering the phoneme /ko/, which consists of a voiceless velar stop and a close-mid back rounded vowel. As a next step in phonation with inhalation, swallowing while breath-holding or phonation of /n/ as an alveolar nasal while elevating the soft palate is effective for achieving glottal airway closure and elevation of the larynx for the second half of insufflation.

Study protocol

Participants sought to learn GPB according to our stepwise protocol. According to the instruction time in our protocol, the maximum time spent in sessions was limited to 60 min for both groups. The DMD group had a maximum of 6 sessions (up to 10 min/session), while the Healthy group participated in a maximum of 4 sessions (up to 15 min/session). In the DMD group, physical and mental stress limited the length of each session. All sessions were conducted within a period of 4 weeks. A participant was considered to have completed the protocol when they had mastered GPB. The following types of feedback were used for GPB learning: tension of the anterior chest as sensory feedback (24, 26, 30, 34); vertical movement of the larynx as tactile feedback with the participant's own hand or visual feedback using a mirror (10); and air leaks from the nose or mouth as visual feedback by nasal mirror or movement of a tissue. In accordance with the protocol, 1 of the 2 trained therapists consistently supervised each participant.

Measurement of vital capacity and glossopharyngeal breathing

Measurement of VC, conducted before each session started, was based on the standard procedure (35). The highest value from 3 trials was recorded (23). The volume of GPB was quantified as VC supplemented by glossopharyngeal insufflation (VC_{GI}), thus VC_{GI}=VC + glossopharyngeal insufflation (16, 24). VC and VC_{GI} measurements were performed by applying a standard oral-nasal mask connected to a Wright Respirometer (DMD group: Wright mark 14; nSpire health, Longmont, CO, USA; Healthy group: SpirobankG; MIR, Rome, Italy) after maximum insufflation or GPB. No nose clip was used in the assessment, because that would not fulfil the purpose of the current study. For efficient learning of GPB, participants started with as many pistons of air as possible, not after the exhalation, but after regular maximal inhalation (24, 34). Boys with DMD self-selected their posture, such as lying supine, sitting cross-legged, or sitting in a wheelchair, while participants in the Healthy group sat on a chair during measurement. Measurements were performed consistently by the therapist who supervised the protocol for each participant.

Outcomes

The primary outcome was the GPB mastery rate based on the VC_{GI}/VC ratio. The VC_{GI}/VC ratio defining GPB mastery was > 1.10 for the DMD group (10) and > 1.05 for the Healthy group (24, 26). In addition, subjective feelings of competence reported by the participant were included as a criterion for GPB. The type of strategies used to achieve GPB was also recorded.

Feasibility outcomes consisted of the number of drop-outs or adverse events, the time required for GPB mastery, and self-reported outcomes (fatigue, difficulty, and motivation). Self-reported outcomes were evaluated by a simple index of fatigue (13, 25, 26), difficulty (23), and motivation (26, 30), which were related to GPB learning. Specifically, subjective parameters before and after daily instruction were recorded on the Borg CR-10 scale (34, 36), which includes fatigue of the respiratory muscles or whole body, difficulty with GPB mastery or instruction, and motivation for GPB mastery. The CR-10 scale of motivation in the healthy group was excluded because it was not a meaningful variable. The main comparison was the change between pre- and post-intervention. Subjective opinions of participants during protocol implementation were also collected.

The results were presented in descriptive statistics. Continuous variables were presented mean (standard deviation (SD)). The feasibility outcomes of CR-10 scale were presented median (interquartile range(IQR)).

RESULTS

Participants

Five of the 25 consecutive participants in the DMD group and 7 participants in the Healthy group were recruited. For the 5 boys with DMD, mean age was 17.8 years (SD 1.3), height 159.6 cm (SD 3.6), weight 55.8 kg (SD 11.4), and VC 1.6 l (SD 0.6). Only 2 patients had used non-invasive positive-pressure ventilation at night and 3 patients had a history of GPB practice with unsuccessful attempts to learn GPB. All patients had taken glucocorticoids. Patients with daytime ventilator use or history of surgery to correct scoliosis were not included in the study in the DMD group. In the Healthy group, mean age was 28.4 years (SD 4.8) , height 168.9 cm (SD 4.2), weight 64.3 kg (SD 9.3), and VC 4.9 l (SD 0.7) .

VC_{GI}/VC and GPB mastery rate

VC_{GI}/VC ratio at the time of GPB mastery and the induction methods that led to mastery by all participants are shown in Table II. For boys with DMD, mean VC_{GI}/VC was 1.25 (SD 0.11). Mean VC at the time of GPB mastery was 1,558.0 ml (SD 332.7), and VC_{GI} increased to 1,970.0 ml (SD 312.6). The induction methods leading to GPB mastery were sucking motion for 3 participants and phonation with inhalation for 2 participants.

In the Healthy group, mean VC_{GI}/VC was 1.09 (SD 0.01) at the time of mastery of GPB. Mean VC at the time of GPB mastery was 4714.3 ml (SD 544.2), and VC_{GI} increased to 5158.6 ml (SD 602.2). There were 2 participants who immediately learned GPB with the imitation method before using the alternative strategies. The alternative induction methods that led to mastery of GPB were sucking motion for one participant and phonation with inhalation for 4 participants.

Feasibility outcomes

The instruction time and self-reported outcomes for all participants are shown in Table II. Since one participant in the DMD group (D1) was difficult to quantify on the CR-10 scale, his subjective parameters were excluded. In the DMD group, mean GPB instruction time was 40.0 (SD 14.1) min (range 30–60 min). No cases showed immediate mastery. No adverse events other than transient fatigue and tension of the anterior chest were seen in any cases. Median fatigue on the CR-10 scale showed a slight increase from 3.5 (IQR 3.0–4.0) at pre-intervention to 5.0 (IQR 1.0–6.0) at post-intervention. Median difficulty decreased from 3.0 (IQR 1.5–5.25) at pre-intervention to 1.0 (IQR 1.0–2.5) at post-intervention. Median motivation increased from 6.5 (IQR 4.5–9.25) at pre-intervention to 9.5 (IQR 7.5–10.0) at post-intervention. In the Healthy group, mean GPB instruction time was 28.6 (SD 13.5) min (range 15–45 min), excluding 2 participants who immediately learned GPB with the imitation method. Median fatigue by the CR-10 scale showed a slight increase from 2.0 (IQR 1.0–6.0) pre-intervention to 2.0 (IQR 1.5–7.0) post-intervention. Median difficulty

Table II. Outcomes at glossopharyngeal breathing (GPB) mastery

				Self-reported outcomes (CR-10 scale)					
				Fatigue (start–end)		Difficulty (start–end)		Motivation (start–end)	
Participants	VC_{GI}/VC ratio	Mastery method	Instruction time, min	Pre-intervention	Post-intervention	Pre-intervention	Post-intervention	Pre-intervention	Post-intervention
D1	1.41	PI	60	–	–	–	–	–	–
D2	1.32	PI	50	4	4	3	1	6	10
D3	1.15	SM	30	3	6	6	1	7	9
D4	1.22	SM	30	3	0	1	1	10	10
D5	1.18	SM	30	4	6	3	3	4	7
H1	1.10	IM	0	2	N/A	2	N/A	N/A	N/A
H2	1.08	IM	0	9	N/A	5	N/A	N/A	N/A
H3	1.09	PI	15	8	1	4	1	N/A	N/A
H4	1.12	SM	45	2	7	4	7	N/A	N/A
H5	1.10	PI	38	2	2	7	2	N/A	N/A
H6	1.09	PI	15	4	7	7	3	N/A	N/A
H7	1.08	PI	30	0	2	4	3	N/A	N/A

VC_{GI}/VC ratio: vital capacity with glossopharyngeal insufflation/vital capacity ratio; D: boy with Duchenne muscular dystrophy; H: healthy men; IM: imitation; PI: phonation with inhalation; SM: sucking motion; N/A: not applicable; –: missing value.

decreased from 4.0 (IQR 4.0–7.0) pre-intervention to 3.0 (IQR 1.5–5.0) post-intervention. All participants in the Healthy group completed the protocol, with no drop-outs. No adverse events other than transient fatigue or chest tension were encountered in any cases.

Participants who could not immediately learn by conventional methods often stated that they understood the mechanism, but imitation of specific movements simply from watching was difficult. For the alternative strategies, they emphasized the benefits of explicit tasks and steps.

Progress for each Duchenne muscular dystrophy case

For all participants, conventional methods had proven difficult to learn, hence application of our protocol was attempted. D number is the serial number of the DMD patient matching Table II.

- *D1.* The inhalation phonation method was selected for practice due to difficulties with sucking motion. To prevent air leaks from the mouth, a bag valve mask with 1-way valve was used in stepwise MIC practice. VC_{GI}/VC increased to 1.41 by session 4 after practice preventing air leaks from the nose. He wished to continue practicing up to session 6 due to feelings of insufficient competence, and more efficient GPB was accomplished.
- *D2.* As sucking motion did not prevent air leaks from the nose and mouth, the inhalation phonation method was used in session 3. To maintain glottic closure, a supervisor encouraged the patient to swallow while breath-holding. To facilitate nasopharyngeal closure, olfactory feedback was provided using detection of aromas. In session 5, VC_{GI}/VC increased to 1.32 along with increased feelings of competence in GPB.
- *D3.* Sucking motion was first performed. To prevent air leaks from the nose, a supervisor provided the patient with olfactory feedback in order to facilitate nasopharyngeal closure. By the end of session 3, VC_{GI}/VC had increased to 1.15, accompanied by feelings of improved competence.
- *D4.* Sucking motion proved difficult to learn. The patient therefore used the inhalation phonation method. However, a supervisor had the patient practice the sucking motion again, in session 3, and this seemed easier at that time. VC_{GI}/VC increased to 1.22, with feelings of improved competence within that third session.
- *D5.* Sucking motion and inhalation method with or without nose clip were attempted, but he failed to achieve mastery because of air leaks from the mouth. Practice of the sucking motion was considered most appropriate given the oral air leaks. The patient practiced this method and achieved a VC_{GI}/VC of 1.18 and increased feelings of competence by the end of session 3.

DISCUSSION

Under the current protocol, all participants in the DMD and Healthy groups were able to master GPB within the allocated 60 min without the use of nose clips. The choice of induction method was highly participant-based. As a result, no single method was applied to all participants. The results suggest that even individuals who had previously been unable to learn GPB were capable of mastering this skill.

The identified feasibility of our methods did not directly verify any specific GPB learning strategies, but instead confirmed the self-reported results. In particular, difficulty and motivation often improved post-intervention, supporting the practicality of the protocol. This is consistent with a previous study showing that learning GPB can be psychologically beneficial (37). No participants showed any adverse events other than fatigue or tension in the chest, all within normal limits. However, most studies including GPB instruction have reported temporary symptoms other than tension in the chest and/or fatigue (16, 24, 26, 30, 34). Shortening the duration of sessions and step-by-step practice in our protocol may have reduced the burden on the participant. The fact that GPB learning was completed within a limited period may be useful in clinical practice, despite GPB learning being recognized as time-consuming.

The rates of mastering GPB can be compared with those of previous studies, revealing rates of approximately 95% in healthy people and athletes (16, 24), 80–100% in patients with spinal cord injury (31, 34), 45.5% in individuals with spinal muscular atrophy type II (30), and 50% in individuals with DMD (10). However, some of those studies allowed the use of nose clips or did not consider GPB self-learning (11, 14, 20, 23, 24, 28, 30, 32). In particular, considering that the previously described learning rate for GPB in patients with DMD was 50% (10), our results suggest that use of a specific protocol might enable more people with DMD to learn this technique. The present methods also seem potentially useful for individuals who encounter difficulties in learning GPB.

After completion of the protocol, VC_{GI} exceeded VC per session in the DMD and Healthy groups by 10% and 25%, respectively. The rate of VC_{GI} increase in previous studies was very high for individuals with long-term use of GPB in cross-sectional surveys (11, 14, 18, 19, 26, 27, 32). In comparison, this rate was 1.12–1.59 for individuals with short-term use of GPB, regardless of medical condition or learning strategy (10, 16, 23, 24, 26, 30, 31, 34, 38). Because continued implementation of GPB could improve VC_{GI} (10, 27), mastering GPB to enable self-training over the long term represents an important first step (34).

The 2 alternative strategies used in this protocol were based on different rationales. Some researchers have shown that movement of the glossopharynx during sucking may be similar to that in GPB (10, 15, 32). In fact, biomechanical analyses of sucking showed similarities to the GPB mechanism of creating differences in air pressure between the oral cavity and pharynx (39, 40). The major difference between GPB and sucking was the presence or absence of nasopharyngeal closure, but GPB seems to be practicable when combined with methods to handle air leak from the nose. On the other hand, phonation methods have been reported in some manuals for induction of appropriate glossopharyngeal movements (13, 23, 25). However, the directness of the method is more important than the appropriate phonation; that is, inhalation rather than exhalation for the desired phonation. The current protocol not only provided guidance regarding proper positioning of the tongue, but also incorporated a phonation method while inhaling to imitate an actual GPB movement. These specific teachings may have reduced subjective difficulty and increased motivation.

The current study was a feasibility case series limited to a small group of practice reports. The primary limitations of this case series were the lack of a follow-up period and the lack of established feasibility outcomes. In addition, monitoring of the volume (in ml) per gulp and gulps per breath, as in the previous study, was not performed. Finally, the DMD group did not include any outpatients or patients with daytime ventilator use. Generalization of the method therefore needs to be performed carefully. Despite the limitations, the present results provide novel insights for clinicians involved in GPB lectures to children with DMD and should be considered for use in future clinical practice. Further research is warranted to validate the current study in a larger cohort.

Conclusion

To our knowledge this is the first study to focus on how to learn GPB in boys with DMD. Sucking and phonation strategies for GPB mastery appear feasible from our limited sample, based on self-reported outcomes, and the lack of either adverse events or drop-outs. For individuals who experience difficulty learning GPB by imitation or from simple instruction, alternative strategies are available that may be worth attempting.

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The authors have no conflicts of interest to declare.

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