



## Early View

Research letter

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# **Glossopharyngeal breathing can allow a lung expansion greater than inspiratory capacity in muscular dystrophy**

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*To the Editor:*

Glossopharyngeal breathing (GPB), also called “frog breathing”, is a positive pressure breathing technique that uses muscles of the mouth and pharynx to propel small volumes of air (“gulps”) into the lungs. It consists of three phases: 1) mouth opening so that air enters the oral and pharyngeal cavities; 2) mouth closure with tongue and pharyngeal muscles propelling a gulp of air through the open glottis, that raises; 3) glottis closure to ensure the air trapped into the lungs[1,2]. These three phases are repeated several times until a comfortable lung expansion is reached. GPB is known to be able to increase lung volumes above physiological total lung capacity (TLC) therefore increasing vital capacity[2–5]. This is the consequence of an increase in lung volume, but also of intrapulmonary gas compression. The physiological factors limiting TLC are the effect of the inspiratory muscles that rapidly decreases and the elastic expiratory (inward) recoil of lungs and chest wall that increase. By contrast, TLC during GPB seems to be limited by sensation rather than by respiratory muscles and/or the mechanics of the respiratory system[3]. GPB is largely used by divers because it increases the amount of oxygen/carbon dioxide stored in the body therefore prolonging their breath-hold duration[3,6,7]. GPB can also be used as lung inflation therapy to assist the ventilatory pump when it becomes weak or impaired, as in the case of cervical spinal cord injury and/or neuromuscular diseases[8].

Muscular dystrophy (MD) is a group of inherited myopathies characterized, among others, by progressive skeletal muscle wasting and weakness, including the respiratory muscles, leading to ventilatory pump failure. In addition, the lung and the chest wall become less distensible, increasing the load against which the ventilator pump must contract. With the progression of MD, a progressive imbalance between the load to be overcome and the capacity to overcome it occurs[9]. Regular mobilization is important to prevent chest wall contractures and/or lung restriction. These patients, therefore, may benefit by GPB as they can recruit the lung, maintain lung and/or chest wall compliance, sustain ventilation for several hours, diminish atelectasis,

improve voice and cough efficacy[2,5,10]. GPB sometimes is learned by the patient himself or it can be trained by specialized physiotherapists. Positive effects of GPB were reported by three authors in patients with Duchenne muscular dystrophy (DMD) in terms of increased peak cough flow[10] and expiratory volume.[2,5]

Measurements during GPB are sometimes difficult to be measured using mouthpiece and pneumotachometer, in particular in patient with macroglossia or severe facial muscles weakness. Lung and chest wall volume variations can alternatively be evaluated by external measurements of chest wall surface motion, like opto-electronic plethysmography (OEP). OEP uses infrared cameras and passive reflective markers on the chest wall, therefore leaving the mouth free. This system also provides the contribution of the ribcage and the abdomen to breathing[11–13].

We used an 8 TV cameras OEP system to measure chest wall and thoraco-abdominal volume variations during quiet breathing, inspiratory capacity (IC) and self-learned GPB in one DMD, one Becker muscular dystrophy (BMD) patient and a competitive breath-hold diver (healthy control, HC) in two postures. The supine position was performed on a rigid support and a 52 markers model was used. In sitting position, the DMD was acquired seated on his wheelchair using the same 52 markers model, whereas an 89 markers model was used for BMD and HC who were seated on the bed without support. All the subjects were asked to reach total lung capacity and then to start GPB until the maximal volume. They were free to control the timing and the amplitude of the manoeuvre according to their sensation.

Forced vital capacity (FVC) and forced expiratory volume in 1 second ( $FEV_1$ ) were measured in sitting position using a spirometer.

The DMD patient was 33 years old, with severe muscular weakness, severe dysphagia, using cough assisted device and continuous non-invasive ventilation. The patient was able to stay disconnected from the ventilator and autonomously breath for short periods of time. During the periods disconnected from the ventilator, his spontaneous breathing was characterized by 3-4 tidal breaths alternated with GPB. Spirometry was unavailable in this patient due to the impossibility to obtain a reliable test because of the severity of the disease, namely facial muscle weakness impeding to hold the mouthpiece with no leakage. He performed the latest spirometry in 2009 (FVC: 0.4 L, 8 %pred;  $FEV_1$  not available). The BMD patient was 19 years old, ambulant, FVC (2.62 L; 55 %pred),  $FEV_1$  (2.10 L; 51 %pred), peak cough flow (287 L/min), not using any

respiratory assistive device. HC was a 29 years old male competitive breath-hold diver, FVC (6.40 L; 106 %pred) and FEV<sub>1</sub> (4.98 L; 107 %pred).

All subjects signed a written informed consent form approved by the local ethical committee according to the declaration of Helsinki. The DMD patient also signed an informed consent for the publication of his video on the supplementary material associated to this research letter.

Because of his age, the specific form of the dystrophy and its progression, the DMD patient was characterized by a severe ventilatory and restrictive pattern. In the DMD and BMD patients as well as in HC, respectively, the following data were measured in the supine position during quiet spontaneous breathing: respiratory rate, 36, 15 and 8 min<sup>-1</sup>; tidal volume, 118, 457 and 930 ml resulting in a rapid and shallow breathing index (RSBI) of 317, 32 and 9 min<sup>-1</sup>·L<sup>-1</sup> (figure 1). RSBI is defined as the ratio of respiratory frequency to tidal volume and it is an important parameter used to predict successful weaning of mechanical ventilation on intensive care units. A respiratory rate of > 38 breaths/min and an RSBI of > 100 breaths/min/L appear to reduce the probability of successful extubation[14].

Respiratory rate (DMD: 29; BMD: 17; HC: 6 min<sup>-1</sup>), tidal volume (DMD: 150; BMD: 618; HC: 1285 ml) and RSBI (DMD: 208; BMD: 30; HC: 5 min<sup>-1</sup>·L<sup>-1</sup>) were measured also in seated position.

In supine position, IC was 0.25, 1.9 and 4.7 litres whereas in seated position it was 0.21, 1.77 and 4.3 L respectively in DMD, BMD and HC. As expected, at the end of the GBP period all the subjects expanded their chest wall well above the volume reached with the IC manoeuvre, the DMD, the BMD and the HC respectively reaching 2.2 L (~880% of IC), 2.5 L (~131% of IC) and 5.4 L (~115% of IC). This was achieved by gulps averaging 45, 40 and 74 ml, respectively in DMD, BMD and HC when lying supine (figure 1). Similarly, in seated position after maximal GPB the volumes reached were ~495%, ~121% and ~113% of IC, respectively in DMD, BMD and HC with gulps of 36, 55 and 77 ml.

GPB has another important effect on abdominal expansion. While ribcage and abdominal volume variations were perfectly in phase in BMD and HC at rest and during IC, they were out of phase in DMD in supine position. For this reason, the contribution of the abdomen in DMD was negative (inspiratory paradox) both at rest (-14%) and during IC (-13%), while it was positive in both BMD (rest: 51%; IC: 42%) and HC (rest: 68%, IC: 37%). Similarly, in seated position inspiratory paradox of the abdominal compartment was still

evident in DMD with a negative contribution both at rest (-3.5%) and during IC (-1.8%), whereas it was still positive in both BMD (rest: 22%; IC: 20%) and HC (rest: 57%, IC: 32%).

During GPB ribcage and abdomen expanded in phase in all subjects (figure 1).

In healthy, during inspiration the diaphragm contracts, it moves caudally with a piston-like movement that makes abdominal pressure and volume increase and pleural pressure become sub-atmospheric to generate inspiratory flow[15]. When the diaphragm is severely weak or paralyzed, as in the DMD patient, the ribcage muscles lead inspiration<sup>12</sup>. Their contraction generates the negative pleural pressure gradient that is partially transmitted to the abdomen because the diaphragm is not able to counteract it. A paradoxical inward movement of the abdominal compartment therefore occurs. The presence of out of phase thoraco-abdominal movement determines an inhomogeneous distribution of lung ventilation, with a reduction of ventilation in the lower parts of the lung due to the cranial shift of the diaphragm during inspiration. By contrast, GPB, driven by oropharyngeal muscles, represents a positive pressure breathing technique that allows to increase, presumably in a stepwise way, alveolar pressure homogeneously in the lung. The expansion of the lower part of the lung is transmitted to the abdomen through a passive diaphragm that finally expands.

We have shown how the volumes change during GPB in competitive breath-hold diver, in a mild and a severe MD patient and that GPB is effective also in presence of severe dysphagia and severe diaphragmatic weakness.

The net effect of GPB seemed milder in the healthy diver compared to MD patients, particularly the most severely compromised one. At the end of GPB, in fact, the increment of volume over IC was substantially greater in MD patients than diver. Although flow and pressure were not measured, it can be speculated that GPB mainly resulted in gas compression in HC and in volume increment in MD. MD patients, although characterized by lung restriction due to the weak respiratory muscles, were able to recruit a substantial additional inspiratory volume by using glossopharyngeal muscles.

The expansion was mainly thoracic in HC and BMD because during GPB, but confined in the abdomen in DMD. This might be explained to be a consequence of the latter to have a more rigid thorax, because of a reduced distensibility of the lung and the stiffer chest wall, while the abdomen became more compliant[9].

GPB not only expands the lung more than IC, but it also reverses paradoxical respiratory abdominal motion with a consequent presumably more homogenous distribution of pulmonary ventilation (i.e., an increased expansion of the lung basis reflecting into a larger diaphragm displacement).

Our data of the DMD patient, in addition, confirmed that GPB is essential for allowing him to stay disconnected from the ventilator, even for short periods of time, and to assist phonation and cough (when cough assisted device was not available) by modulating the amplitude of the inspiratory volume.

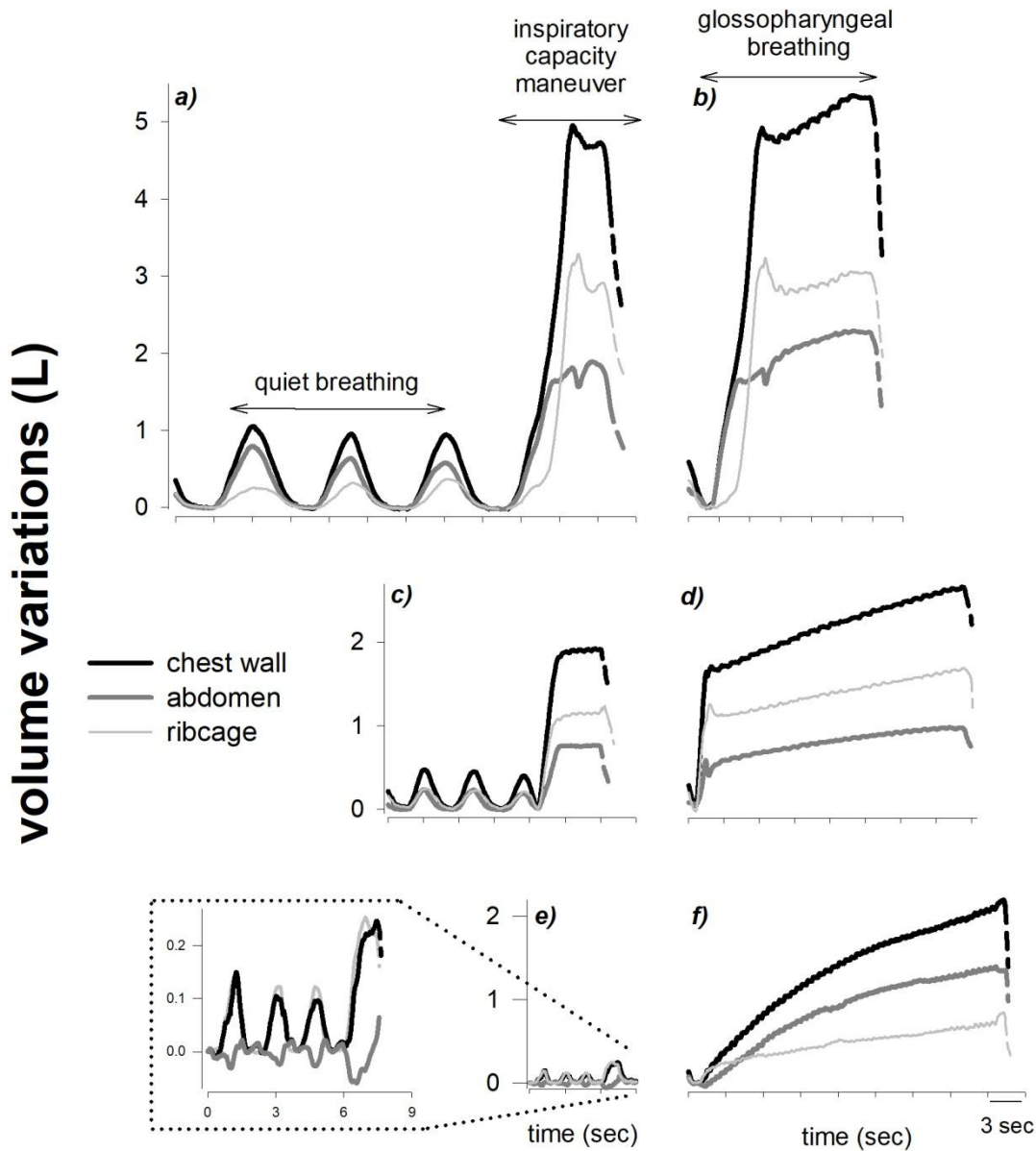
For all these reasons, our data suggest that patients with intact bulbar function, even in presence of muscular weakness/atrophy/paralysis, should be trained and/or encouraged to perform GPB daily.

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**Figure legend:** Volume variations of the ribcage (grey line), the abdomen (thick grey line) and the chest wall (thin black line) during three breaths followed by an inspiratory capacity (a, c and e) and during glossopharyngeal breathing (b, d and f) in a competitive breath-hold diver (a and b), a Becker muscular dystrophy (c and d) and a Duchenne muscular dystrophy (e and f) patients.



**Supplementary material legend:** Recording of DMD patient during quite breathing, inspiratory capacity and glossopharyngeal breathing in supine position (right), corresponding 3D markers movement (top left) and ribcage (blue), abdomen (red) and chest wall (grey) volume traces (bottom left).