More than good lungs!

Mais do que bons pulmões!

Ilma Aparecida Paschoal

Gas exchange requires more than two reasonably normal lungs, and neuromuscular diseases illustrate quite well the concept expressed by this statement.

Knowledge of respiratory physiology increased significantly during periods of crisis in the world. The poliomyelitis pandemic that occurred between 1900 and 1950 is an example of progress achieved through pressing need. The pandemic showed that it was vital to have machines that would be able to substitute for spontaneous ventilation. However, interest in diseases that interfere with alveolar air exchange through muscular or neurological deficits, or a combination of the two, seems to have dwindled in recent decades.

Our specialty in particular has not paid due attention to these patients, although noninvasive pressure support ventilation is increasingly available in Brazil.

American physician John R. Bach has extensive experience in treating these patients. Although he is a neurologist, Dr. Bach devotes a great deal of attention to respiratory impairment in neuromuscular diseases, since respiratory complications in these individuals are the major cause of morbidity and mortality. Competent and charismatic, Dr. Bach applies various techniques for respiratory support and for preventing the accumulation of secretions; he has demonstrated the effectiveness of these techniques in various studies.[1-3] He is the author of the aphorism that states that patients with neuromuscular diseases die not because they cannot breathe, but because they cannot cough properly.

In an editorial published in 2003,[4] Bach stated that he had the opportunity of clinically monitoring more than 700 patients with different neuromuscular diseases, such as post-polio syndrome, spinal trauma and other conditions in which there is respiratory muscle weakness or paralysis. These individuals were treated with noninvasive positive pressure ventilation, delivered continuously through mouthpieces or nasal masks (or a combination of the two) sometimes for quite long periods of time, as long as 40 years. However, it would have been absolutely impossible to avoid progression to a tracheostomy if effective airway clearance had not been performed in cases of accumulation of secretions.

Mucociliary transport in nontracheostomized patients with neuromuscular disease is normal. Cough, however, is not.

For cough to occur, inspiratory muscles must provide pulmonary inflation of up to 85-90% of total lung capacity. Firm glottal closure, for approximately 0.2 s, follows. Vocal fold movement requires that the laryngeal muscles innervated by bulbar neurons contract. The subsequent contraction of the expiratory muscles (intercostal and abdominal muscles) produces intrapleural pressures as high as 140 mmHg. The sudden opening of the glottis after the contraction of the expiratory muscles generates peak cough flows (PCFs) that range from 360 to 1,200 L/min; these PCFs are facilitated by gradual vocal fold abduction.[5] Therefore, cough depends on the preservation of inspiratory, expiratory and bulbar muscle functions.

The expiratory volume during cough is 2.3 ± 0.5 L. A tidal volume of at least 1.5 L must be previously inhaled for minimally effective cough to be achieved.[6]

In order to achieve an adequate PCF in patients with low FVC, it is crucial that deep insufflation (through the ventilator being used) or air stacking be performed.

Air stacking allows the accumulation of sufficient volume in the lungs to achieve an acceptable PCF. Air stacking begins with a deep inhalation (spontaneous or assisted by a ventilator or by a manual resuscitator). The patient then receives repeated volumes from the ventilator or manual resuscitator. The glottis opens to receive the new volume and soon closes. If the cheeks or lips are too weak to allow air stacking to be performed, the maneuver is performed through an oronasal or nasal mask.[4] The ability to perform air stacking indicates the degree of

preservation of bulbar muscles.[7] Great volumes of air can be stored in the lungs under pressures ranging from 40 to 70 cmH_2O, and sudden exhalation of this air, assisted by chest or abdominal compression (or a combination of the two), can adequately substitute for coughing. It is estimated that a PCF of at least 160 L/min is needed in order to clear central airway secretions.

Air stacking as part of the assisted cough technique effectively achieves sufficient PCFs to improve clearance of airway secretions in patients with severe impairment of inspiratory and expiratory muscles,[1-3] a fact that was also noted by Brito et al., in a study published in this issue of the Brazilian Journal of Pulmonology.[8]

In most patients with neuromuscular diseases (with the exception of very young children, who are unable to cooperate, and patients with amyotrophic lateral sclerosis and bulbar muscle involvement), the maneuvers that aim to increase PCFs can achieve values higher than 160 L/min. Complete failure of the muscles of the larynx, which makes vocal fold movement impossible, can render even the cough machine (Cough Assist; Philips Respironics, Murrysville, PA, USA) ineffective, since the trachea tends to collapse during inhalation and exhalation in this situation. In this phase of the disease, only a tracheostomy can prevent respiratory failure.[4]

Therefore, it is of utmost importance that, in patients with neuromuscular diseases, the effectiveness of cough be evaluated by measuring PCF. If cough proves ineffective, cough assist techniques are mandatory, especially during episodes of respiratory infections (even the most banal ones, such as colds). The air stacking technique is quite effective, and its application should be popularized in Brazil. The experiment conducted by the aforementioned authors and reported in this issue of our Journal addresses this topic.

Ilma Aparecida Paschoal
Associate Professor.
Department of Clinical Medicine,
School of Medical Sciences,
***Universidade Estadual de Campinas* –**
Unicamp,
State University at Campinas –
Campinas, Brazil

References

1. Bach JR, Alba AS, Saporito LR. Intermittent positive pressure ventilation via the mouth as an alternative to tracheostomy for 257 ventilator users. Chest. 1993;103(1):174-82.
2. Bach JR. Amyotrophic lateral sclerosis: prolongation of life by noninvasive respiratory aids. Chest. 2002;122(1):92-8.
3. Gomez-Merino E, Bach JR. Duchenne muscular dystrophy: prolongation of life by noninvasive ventilation and mechanically assisted coughing. Am J Phys Med Rehabil. 2002;81(6):411-5.
4. Bach JR. Mechanical insufflation/exsufflation: has it come of age? A commentary. Eur Respir J. 2003;21(3):385-6. Comment on: Eur Respir J. 2003;21(3):502-8.
5. Leith DE. Cough. In: Brain JD, Proctor D, Reid L, editors. Lung Biology in Health and Disease: Respiratory Defense Mechanisms, Part 2. New York: Marcel Dekker; 1977. p. 545-92.
6. Bach JR. Mechanical insufflation-exsufflation. Comparison of peak expiratory flows with manually assisted and unassisted coughing techniques. Chest. 1993;104(5):1553-62.
7. Kang SW, Bach JR. Maximum insufflation capacity. Chest. 2000;118(1):61-5.
8. Brito MF, Moreira GA, Pradella-Hallinan M, Tufik S. Air stacking and chest compression increase peak cough flow in patients with Duchenne muscular dystrophy. J Bras Pneumol. 2009;35(10):973-9.