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Letters to the Editor

Colonic distension treatment in Duchenne muscular dystrophy

We read with interest Vianello's work [1] and we think that this is an important event during noninvasive ventilation (NIV) in neuromuscular disease especially in dystrophy. However, we want to draw attention to some considerations for proper practical applications:

First, dystrophin is expressed in the smooth muscle of the gastrointestinal tract. Normally, a disruption of protein expression can lead to functional disturbances of the gastrointestinal tract including acute gastric dilatation, gastroparesis and intestinal pseudo-obstruction. DMD patients with advancing age may suffer from nutritional problems due to swallowing impairment, collection of gastric air, gastro-esophageal reflux and chronic constipation that may lead to life threatening complications [2].

The impaired gastrointestinal function may further complicate the care of older DMD patients who already suffer from progressive impairment due to general loss of muscular strength, respiratory and cardiac failure. There is a significant risk of Duchenne patients developing life-threatening constipation in combination with metabolic acidosis when progressive difficulty in swallowing leads to insufficient fluid and caloric intake and with further impairment of gastrointestinal motility [3].

Importantly, Lo Cascio et al. found that symptoms of impaired gastrointestinal function were not significantly correlated with objective measures of impaired gastrointestinal transport, suggesting that patients may not perceive potentially dangerous constipation as requiring treatment.

Second, aerophagia is an important NIV-related problem and it appears in up to half the patients with NIV and may lead to discontinuing treatment (Fig. 1) [4].

Gastric distention compresses the lungs and induces high intrathoracic pressures that therefore require high inspiratory pressure to ventilate the lung. This generates more air leak, and the ventilator compensates by increasing inspiratory flow and thereby creating a vicious circle.

Treatment of abdominal distension includes drugs to accelerate intestinal transit and adjusting the ventilatory settings. Reducing the volume released by the respirator can relieve the patient's discomfort, though at the cost of using lower insufflation pressure resulting in a certain loss of ventilatory efficiency. Peak pressure can also be regulated by increasing the pressure ramp slope on ventilators equipped with that function or by adjusting the inspiratory to expiratory flow rate. It is also important to regulate the inspiratory and expiratory trigger for better patient ventilator synchronization. Alternating ventilatory modalities (pressure and volume) can also help to correct problems of aerophagia. Finally, it has been documented that the problem may disappear after a few weeks of treatment, either spontaneously or because the patient has learned to handle and eliminate intestinal gas more effectively [3].

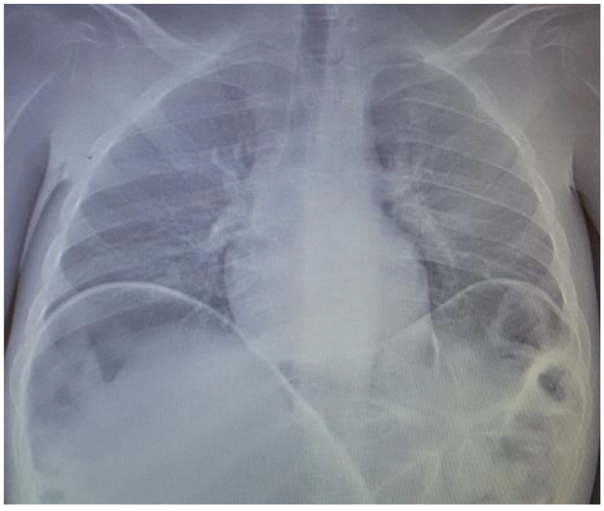

Fig. 1. Dilated bowel loops and upward displacement of the diaphragm in a DMD patient's chest X-ray.

The air usually passes as flatus when the patient is mobilized in the morning. If severe, it can increase ventilator dependence and result in the need for a gastrostomy or nasogastric tube to burp out the air, or a rectal tube to decompress the colon. If these measures do not correct the problem, it is reasonable to rule out gastric disease before considering withdrawing the respirator or performing a tracheostomy in order to switch to invasive ventilation.

Consequently, we must exercise caution with patients we start on NIV who experience aerophagia that does not respond to the usual therapeutic measures. We must consider exploration of the digestive tract to be a good practical

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option before considering other ventilatory alternatives or even discontinuing noninvasive ventilation altogether.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.nmd.2018.10.004.

References

[1] Vianello A, Arcaro G, Peditto P, Iovino S, Marchese-Ragona R. Acute colonic distension in a patient with severe muscular dystrophy receiving non-invasive positive pressure ventilation. Neuromuscul Disord 2018;28(1):60–1.

[2] Lo Cascio CM, Goetze O, Latshang TD, Bluemel S, Frauenfelder T, Bloch KE. Gastrointestinal dysfunction in patients with Duchenne muscular dystrophy. PLoS One 2016;11(10):e0163779. doi:10.1371/journal.pone.0163779.

[3] Lo Cascio CM, Latshang TD, Kohler M, Fehr T, Bloch KE. Severe metabolic acidosis in adult patients with Duchenne muscular dystrophy. Respiration 2014;87(6):499–503.

[4] Fiorentino G, Annunziata A, Cauteruccio R, Frega GS, Esquinas A. Mouthpiece ventilation in Duchenne muscular dystrophy: a rescue strategy for noncompliant patients. J Bras Pneumol 2016;42(6):453–6.

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