Pediatric Pulmonology 49:1045–1046 (2014)

Response to Letter

Methodological considerations in combined noninvasive ventilation and mechanical in-exsufflator

Dear Editor,

It is our pleasure to read the comments sent to the journal by Equinas et al. regarding our article "Combined noninvasive ventilation and mechanical in-exsufflator in the treatment of pediatric acute neuromuscular respiratory failure."[1] The exciting field of critical care of acute respiratory failure (ARF) in pediatric patients with neuromuscular diseases (NMD) is clearly challenging because of its unique, heterogeneous, and vulnerable features. Thus, we are glad that other centers are interested in these populations. We thank Equinas et al. for their valuable comments, which enabled us to better address some important issues in our article.

To date methodological data regarding combined use of noninvasive ventilation (NIV) and mechanical in-exsufflator (MIE) are limited, especially in the setting of the pediatric intensive care unit (PICU). Even though the present study is the largest cohort to date in neuromuscular patients with ARF in PICU, we agree that its sample size is small compared with most reports in adults, with dispersed age distribution and heterogeneous NMD. However, these features are common in most pediatric studies of NMD, reflecting their relatively low incidence in general populations. We also thank that Equinas et al. discussed about the prevalence of bulbar weakness, which raised an important point. It has been well known that bulbar weakness may lead to the risk of a aspiration of food and saliva, raising the risk of NIV failure,[2] such as in patients of spinal muscular atrophy type 1 (SMA1) or bulbar amyotrophic lateral sclerosis. Therefore, in our study, we have excluded patients with profound bulbar weakness. However, we did enroll patients with SMA1, because all of them had received gastrostomy and Nissen fundoplication to avoid potential respiratory insults caused by reflux, as suggested by a recent guideline for standard care of SMA.[3]

We agree that lung function test (LFT) may be an objective tool to assess the responses to interventions in stable patients with respiratory insufficiency. However, we did not perform LFT in the present study due to several reasons. First, LFT is frequently unpractical in children with NMD, as a previous study has demonstrated that even in stable children forced vital capacity could not be obtained in 40% of them.[4] Indeed, a recent guideline of respiratory care for children with NMD established by the British Thoracic Society recommends the measurement of peak cough flow only in children older than 12 years old.[5] In addition, in the setting of PICU, and for infants like those with SMA1, LFT may be difficult or impossible. Thus, in our experience we believe that, unlike the role of LFT in adults or stable older children, clinical parameters such as blood gas analysis, respiratory rate, and heart rate may be more practical to assess the acute changes of respiratory status and the effective clearance of secretions after NIV/MIE interventions in critically ill children with acute pulmonary infections.

Hypercapnic respiratory failure is common in patients with NMD as shown in our study. Equinas et al. are also concerned about the hypercapnia in the NIV/MIE failure group in our study. However, since there was also hypercapnia in the successfully treated group and there was no statistically significant difference between them, we believe that it is not a risk factor for intubation in the present study.

Sedation is a major issue in methodological considerations of NIV. We understand that most intensivists are reluctant to use sedation and analgesics for patients on NIV due to concern of respiratory drive depression.[6] However, our experiences suggest that mild sedation in infants and younger children may improve patient–ventilator

Conflict of interest: None.

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Received 4 October 2013; Accepted 8 November 2013.

DOI 10.1002/ppul.22964
Published online 23 December 2013 in Wiley Online Library (wileyonlinelibrary.com).

synchrony, which has been demonstrated as a positive factor for NIV success.[7] In fact, it was found that up to 14% of patients receiving NIV in ARF were unable to tolerate face-mask ventilation.[8] In another study, Antonelli et al.[9] showed that mask intolerance or inadequate patient cooperation led to intubation in 9% of patients with ARF. Thus, in our study, mild conscience sedation was achieved by low-dose chloral hydrate or midazolam, similar to a recent pediatric NIV study,[10] which did not report any adverse cardiopulmonary events due to these sedatives.

All of our patients except patient No. 7 had hypercarbic respiratory failure, as this is very common in patients with NMD. In fact, in the setting of PICU or emergency department, hypoxemia in these patients is frequently caused by inadequate clearance of secretions, hypoventilation, and atelectasis. In this context, the goal of treatment is to normalize gas exchange by delivering positive airway pressure to decrease atelectasis and to optimize airway clearance, while supplemental oxygen is not the first line of treatment. Supplemental oxygen may result in improved oxygen saturation, but may also mask hypoventilation or impair central respiratory drive and does not improve ventilation or treat the underlying causes of hypoxemia in these patients with failure of respiratory pump.[11]

Finally, we agree that current data regarding optimal mode of ventilatory support are still very scarce and this issue should be addressed in multicenter large-scale trials since combined NIV/MIE appears to be a promising strategy in the management of ARF and prevention of extubation failure, even in the general population without NMD.[12] Most importantly, a need is evident for more sophisticated clinical and physiologic parameters to assess responses to this noninvasive approach. These should be tailored to different age groups and to different NMD with varying degrees of respiratory muscle involvement.

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