



## COMMENT

# Mechanically Assisted Invasive Ventilation for ALS patients: Is it the Ultimate Strategy To Improve Survival?

## A Ventilação Mecânica Invasiva na Esclerose Lateral Amiotrófica: a derradeira estratégia?

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Ambrosino and his colleagues from an Italian center at Pisa, describe their experience with a significant impact on the management of home-ventilated ALS patients. This work certainly opens a new era by encouraging new studies on clinical evolution and prognosis, thereby deemed to reduce the uncertainties experienced for all of those responsible for the follow-up of these patients that may improve the palliative care at the end-of-life.

Recent evidence aggregating results from retrospective analysis of autopsies of ALS patients with their respective reported clinical causes, showed that despite the frequent lack of concordance, they found that respiratory infectious conditions were the most prominent features and that pulmonary embolism was observed only in spinal ALS patients, and heart failure as unexplained death was more than two-fold higher in bulbar ALS patients than in the spinal forms.<sup>1</sup>

In addition, bulbar ALS patients have more frequently disautonomy and sympathetic hyperactivity, and specifically in ventilated ALS patients these phenomena have already been for many years described and recognized as possible cause of unexpected death among ventilated patients.<sup>2</sup>

Moreover, it is recognized that diffuse alveolar hemorrhage is also an unspecific feature, both for ARDS with mechanically lung injury produced with higher lung volume, and bronchopneumonia. Indeed, Prost and colleagues found

strong and powerful associations between those clinical conditions in multivariate analysis.<sup>3</sup>

However, as Diaz and colleagues, reported that diffuse alveolar hemorrhage is the clinical feature that often may not be apparent or its presentation can be mistaken for an alternative disease process, and may be completely unrecognized even in ICU.<sup>4</sup>

Accordingly, we secured it to the so called “irrelevant data” on the 2 previous admissions to the emergency room in the last four months described in the text by Ambrosino and colleagues, to draw attention and focus to this critical condition in the emergency room.

Indeed, the presence of an obvious critically ill ALS patient like the one reported on this clinical case, probably with an end-stage disease, the diffuse alveolar hemorrhage was very likely the cause of death. Nevertheless, the etiology attributed to malfunction of the device without discussion of other clinical conditions seemed rather partial especially because of the high frequency of other possible negative events in ALS patients.

Still, it is not disputable the interest of the present case study and future research should cover these pertinent issues, as it is granted that other causes of death are implicated in the terminal ALS stages, even if properly and adequately invasively ventilated.

Overall, though mechanically assisted invasive ventilation (MAIV) is often believed to be a life-prolonging intervention, it is not devoid of serious life-threatening complications, and the long-term medical course of ALS with MAIV has never been tested in a randomized trial with NIV

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that frequently goes up to 24/24 and also to the point of total locked-in syndrome, meaning that, especially due to the paucity of studies, we may no longer consider MAIV as the ultimate strategy to improve survival but rather it should be more extensively evaluated.

On the other hand, as the risk management and safety of the home ventilator dependent patient is impossible to guarantee<sup>5</sup> especially care has to be undertaken by manufacturers when implementing software designs to detect and anticipate possible faulty mechanisms. They should be readily accessible by physicians, providers, patients and carers, and assist immediate responses. However, even in the last generation of ventilators the alarm signs are not easily transmitted, mostly due to the protection of data laws that do not allow a good communication with carers and patients which, once fully educated and familiarized with problems and solutions should be thought as the true experts and team members.

In conclusion, though the authors could have reported a more detailed clinical case and improved the discussion, this report is very interesting and quite important and possibly may represent a turning point, calling for more detailed studies on durability, reviews and

maintenance of devices as well as in imposed demands both on manufacturers and providers and regulatory mechanisms regarding patient safety.

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