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dying

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EDITORIAL

ALS: Control ventilation, manage respiratory secretions and, when required, oversee the process of dying



ELA: controlar a ventilação, manobrar as secreções respiratórias e, quando necessário, supervisionar o processo da morte

ALS is a neurodegenerative disease which leads to progressive paralysis. While there is no cure for ALS, treatment is available and, as recently said to us a board member of a Spanish ALS patient association, “our doctor is the pulmonologist”. This is true, but only to an extent. The contribution of a neurologist and other professionals is essential for the management of some of the problems which damage quality of life of patients and their relatives.¹ Moreover, respiratory care teams are committed to move treatment away from the environment of palliative care, for those patients who desire this, and move it towards what A. Oppenheimer called “high technology and compassionate care.” Yet, we must also prevent dyspnoea and any other kind of suffering for those patients who refuse life-sustaining treatment.

The old statistics on the survival of ALS patients are no longer valid because their lives can now be prolonged – without damaging quality of life and even improving it – thanks to the respiratory muscle aids now in use: aids for the maintenance of adequate alveolar ventilation and the appropriate management of pulmonary secretions. Non-invasive procedures have demonstrated their ability to have a positive effect on the prognosis of these patients and they constitute extremely valuable tools until the impairment of the bulbar-innervated muscles becomes so severe that they become ineffective.^{2,3} Thus, the muscles which cause non-invasive ventilation techniques to fail are not the thoracic muscles (as inexperienced doctors might suspect) but those which are bulbar-innervated. Patients can be kept alive by

means of non-invasive techniques without the patient being required to make any effort to breathe or cough,^{2–4} even during episodes of pneumonia⁵ – bulbar impairment permitting.

To achieve this, comprehensive knowledge of the available tools and their use is necessary: the right ventilators, masks of various types, procedures for cough capacity assessment and coughing aids where necessary, having in mind that, as Dr Escarabill wrote,⁶ “the therapy “package” must also include the organisation of care”. A good doctor must not introduce devices too early into the home of a patient nor delay this unnecessarily; patients must be carefully studied in order to ensure that they receive the most effective and efficient treatment. And let us not forget that very few patients refuse non-invasive procedures, whereas the percentage of patients who refuse invasive treatment is relatively high.⁴ Therefore, when we accept the responsibility to take on the care of a patient, it also becomes our responsibility to be masters of the techniques which can provide support to the respiratory muscles. When speaking of such expertise, it is important to remember that what is of real value in a unit for ALS patients is not the fact that the patient may be visited by a range of specialists in the course of one day. Of course, such co-ordination is of interest, but it becomes of little value if the shift schedule is such that these patients are at times left in the care of doctors with little or no expertise in non-invasive ventilation or who are unaware of how to undertake patient-centred, informed decision-making. Can anyone imagine a lung transplant patient being treated in an emergency by a doctor with no relevant experience?

When non-invasive procedures fail, there are two alternatives, which the patients and their family members must have been able to consider previously in good time, having received full information on them: tracheotomy ventilation

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and comprehensive palliative care.^{1,4,7} Units specialising in the care of ALS patients must be able to provide them with both of these options at excellent standards of quality. Doctors should not begin treatment involving mechanical ventilation if they cannot then end such treatment in the proper manner, if this is the patient's wish.⁸ We must minimise the suffering of patients and relatives.

This edition of the Rev Port Pneumol makes a significant contribution to knowledge regarding the treatment of respiratory problems suffered by ALS patients thanks to the work of a group of experts from across Europe.^{6,9,10} However, we must also pay tribute to the work of someone who for many of us has been the real pioneer. He was the one who enabled us to change our procedures and multiply our therapeutic effectiveness while minimising treatment side-effects. We have come a long way on our own now, but we must ensure that those new to the field understand that it was JR Bach who showed us the way.

In order to improve treatment, in the early 2000s we would often stress the importance for the presence of units specialising in the respiratory care of ALS patients.¹¹ Unfortunately, until very recently, the lack of dedicated resources for such patients was still a common complaint, as their rights to be treated fairly were clearly not being respected.¹ However, things are changing. The profile of ALS patients is rising in both medical and social contexts and pulmonologists are showing particular interest in acquiring the right knowledge and skills to be able to provide them with high-quality care. This journal is an example of this change.

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