The progression of amyotrophic lateral sclerosis: respiratory function

To the Editor: We would like to offer a few comments on the article by Salord et al[1], which we feel to be of great interest. The authors report the evolution of respiratory function in a 26-year-old patient with amyotrophic lateral sclerosis (ALS) from the time that he is diagnosed until his death from respiratory failure. For a period of just over five years they monitor the gradual reduction of spirometric parameters, maximal respiratory pressures and maximum voluntary ventilation. They also highlight the appearance of a pattern of instability in the upper respiratory tract 18 months into the illness. At the end of the article the authors suggest that these values should be included among the variables regularly used to follow the progression of this illness. However, we were surprised by the absence of any reference to certain aspects that we consider to be fundamental in monitoring the progress of these patients.

1. There is a surprising lack of data related to pulmonary gas exchange, which we think are very important in monitoring the progress of patients with ALS. We are referring in particular to arterial blood gas values and nocturnal pulse oximetry readings. According to the scientific evidence at our disposal, it is advisable to initiate the use of noninvasive ventilatory support systems when $PaCO_2$ is greater than 45 mmHg, when the nocturnal pulse oximeter shows oxygen saturation levels of less than 88% over 5 consecutive minutes, when forced vital capacity (FVC) is less than 50%, or when the maximal inspiratory muscle pressure is less than 60 cm H_2O[2]. The lack of information on these aspects prevents us from knowing how long the patient suffered from respiratory insufficiency before he died and whether he received any kind of ventilatory support.

2. According to the available data on the progression of respiratory function in the patient, vital capacity was 43% and maximal inspiratory pressure 44 cm H_2O in September 1993, nearly three years before death. According to the criteria mentioned above, he should then have received noninvasive mechanical ventilation (NIV)[3]. The authors should have made some mention of this stating whether or not they started mechanical ventilation and if so what effect it had on the patient's evolution. They should also have mentioned whether they considered providing a tracheostomy at any point.

3. The pathophysiology of respiratory failure in patients suffering from neuromuscular diseases is a complex subject. It may include changes in the mechanical properties of the respiratory system, the appearance of signs of muscle fatigue, changes in respiratory control, changes in nocturnal pulmonary gas exchange causing decreased sensitivity of both central and peripheral chemical receptors, and upper respiratory tract dysfunction[4]. Nocturnal respiratory support has proved to be effective in correcting daytime respiratory failure in these patients, probably because it has a greater or lesser effect on the diverse mechanisms involved. This is why it is important to evaluate all aspects of a patient's pulmonary function and not just the spirometric parameters. In the absence of other data indicating the need to initiate NIV, it may make sense to use spirometric parameters to detect a reduction of FVC to 50% in time, but it makes no sense after this point.

4. As the authors say, ALS can progress at different rates. As well as the slowly progressive forms of the illness, we sometimes see forms that start with acute respiratory failure. For these patients diagnosis takes place when the patient has already undergone a tracheostomy and is in an intensive care unit[5,6]. On other occasions bulbar symptoms predominate. In many cases it is impossible to carry out spirometric measurements and the only techniques available for evaluating the patient's respiratory capacity are pulse oximetry and arterial blood gasometry. We find it difficult

to understand the need for recording the spirometric measurements of a patient every 3-4 months until the time of death.

To sum up, we think that the assessment of ALS patients should be a comprehensive process in which spirometric parameters and maximal muscle pressures assessed at regular intervals do not necessarily play the most important part. Perhaps we should give priority to assessing the right moment for starting mechanical ventilation, to improving the mechanisms for effective coughing, to providing patients and families with reliable and complete information on the progression of the illness and its consequences, and to obtaining informed consent if the time comes to decide whether to create a tracheostomy or simply to accept a dignified death.

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