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Early Non-Invasive Ventilation in Hypercapnic Respiratory Failure: A Case of Undiagnosed ALS

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Introduction

It is difficult to detect hypercapnic respiratory failure caused by restrictive pattern pulmonary diseases in neuromuscular diseases using imaging evaluation or pulmonary function tests, which are generally performed in the respiratory department. When such respiratory failure occurs, it is common to intubate the patient, treat them in the intensive care unit, and then perform multiple weaning attempts before performing a tracheostomy and using a full-time ventilator. This is a case in which hypercapnic respiratory failure that occurred before the diagnosis of amyotrophic lateral sclerosis (ALS) was treated by applying prompt non-invasive ventilation (NIV) even in an emergency situation of respiratory failure after detailed pulmonary evaluation using different evaluation methods from those used in the Department of Respiratory Medicine in the Department of Rehabilitation Medicine.

Case presentation

In September 2022, a 46-year-old man presented with right upper limb numbness and weakness. Cervical MRI showed spinal stenosis at C5–6 with myelopathic signs (positive Hoffman, clonus, Babinski), leading to a diagnosis of cervical spondylotic myelopathy. He underwent Anterior Cervical Discectomy and Fusion operation.

One month postoperatively, despite improved limb strength and stable radiographs, he complained of significant dyspnea even with minimal exertion. At the 3-month follow-up, his dyspnea persisted, prompting a respiratory consultation. In December 2022, after a COVID-19 infection, a pulmonary function test demonstrated a restrictive pattern with a sitting vital capacity (VC) of 1.9 L (42% predicted); chest CT scans were unremarkable.

In February 2023, the patient visited outpatient clinic of Department of Physical Medicine and Rehabilitation where his evaluation revealed, **severely reduced supine VC 0.98 L (23%), and maximal inspiratory pressure (MIP) 14 cmH_2O (15%), maximal expiratory pressure (MEP) 22 cmH_2O (16%), SpO_2 of 86% at rest and 77% during walking**. Given these findings, he was immediately transferred to the ER, where arterial blood gas analysis showed ***severe hypercapnic respiratory failure (pH 7.257, pCO_2 105 mmHg, pO_2 60.8 mmHg, SaO_2 87.7%)***. Since he remained alert and cooperative, NIV was initiated using a portable ventilator and ambu bag without performing intubation. Over several hours his blood gases improved and he was moved to the E-ICU on next day.

The following day, he was transferred to the general ward and gradually weaned from NIV. By day 5, he was discharged with a portable ventilator—using NIV during sleep and intermittently during the day—which enabled him to return to work (Table1). Follow-up tests initially showed improvement in VC, MIP, and MEP; however, these parameters later declined, and two years after respiratory failure onset (Figure 1). His pulmonary function deteriorated nearly to initial levels, with increased daytime ventilatory support. He was eventually diagnosed with ALS; by fall 2024, worsening lower limb weakness led to wheelchair dependence and declining upper limb strength resulted in work leave.

Date/Time	pH	pCO_2 (mmHg)	pO_2 (mmHg)	O_2 Sat (%)	Major event and interventions
2022-09-06 (Preoperative Admission)	7.35	55 ↑	85	95	Preoperative evaluation – mild hypercapnia detected
2022-09-07 (Surgery Day, ACDF C5-6)	7.32	65 ↑	75	93	ACDF C5-6 performed – hypercapnia (+)
2022-09-14 (Orthopedic Discharge)	7.40	47 ↑	90	96	Postoperative improvement – reduced pCO_2, improved ventilation, discharged
2023-02-15 (Outpatient Clinic, PM&R OPD)	–	–	–	85 (resting), 77 (walking)	Rehabilitation outpatient evaluation – Severe oxygen desaturation detected via SpO_2, transfer to ER decided
2023-02-15 15:26 (ER Arrival)	**7.20 ↓**	**110 ↑**	**55 ↓**	**85 ↓**	Initial ABGA in ER – severe hypercapnia and hypoxemia confirmed
2023-02-15 15:53 (ER)	**7.257 ↓**	**105 ↑**	**60.8 ↓**	**87.7 ↓**	Persistent acidosis and hypercapnia detected in first ABGA
2023-02-15 16:18 (ER)	**7.300 ↓**	**98 ↑**	**65**	**88.5 ↓**	Before NIV application – slight improvement, mild decrease in pCO_2, slight increase in pO_2 and O_2 Sat
2023-02-15 16:37 (ER)	7.450	**80 ↑**	75	93.0	NIV treatment initiated – pCO_2 decreasing further, O_2 saturation improving
2023-02-15 17:32 (ER)	7.580	**55 ↑**	95	96.0	After NIV application – gradual respiratory stabilization
2023-02-15 19:13 (ER/E-ICU)	7.580	41	110	98.9	Final ABGA with NIV – pCO_2 normalized, O_2 saturation fully recovered
2023-02-16 12:20 (General Ward Transfer)	–	–	–	–	Condition stabilized, transferred to general ward, partial NIV weaning initiated
2023-02-16 20:42 (General Ward)	7.597	27.5	109	99.3	Condition stabilized, partial NIV weaning continued, pCO_2 normalized, stable respiratory status. Oral feeding resumed.
2023-02-19 06:37 (Pre-Discharge Evaluation)	7.418	49.7	81.2	97.1	Final ABGA – off NIV during daytime, recovered to functional activity level
2023-02-19 (Discharge)	–	–	–	–	Home NIV prescribed – mostly off NIV during the day, returned to normal work life

Table 1. Timeline of hypercapnic respiratory failure events and pulmonary rehabilitation interventions

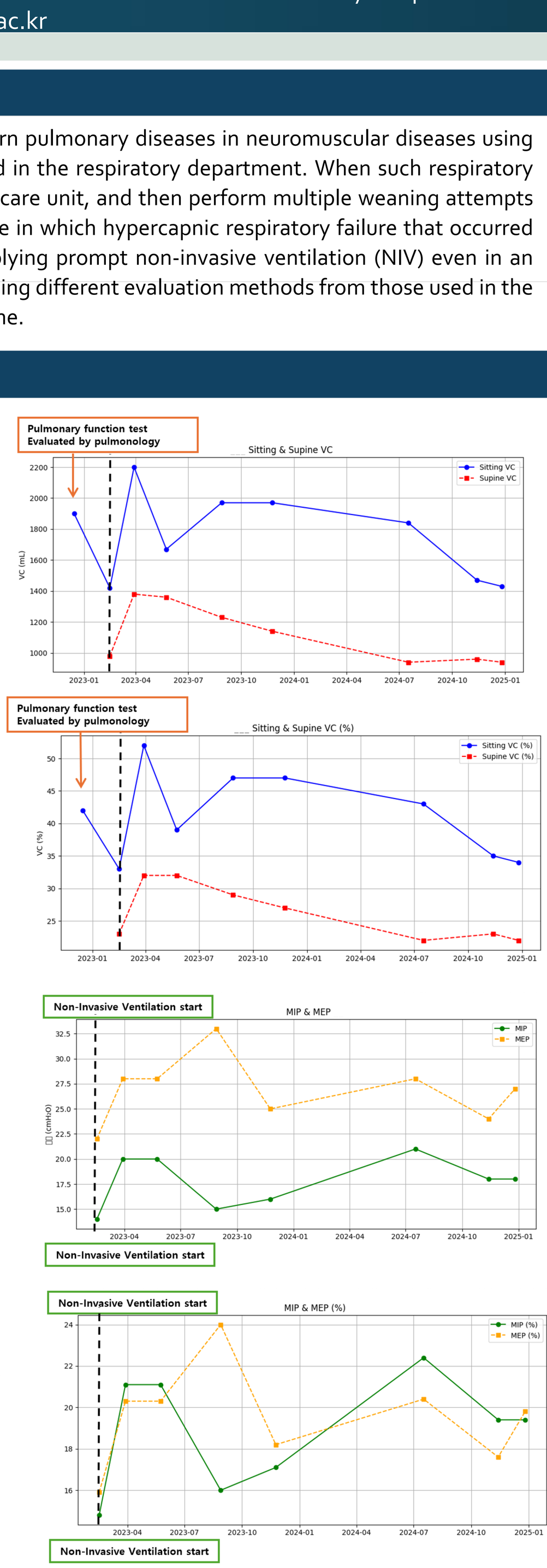

Figure 1. Timeline of pulmonary function test in Respiratory-Onset ALS Patients
At the time of hypercapnic respiratory failure, the patient showed severely reduced pulmonary function, with a supine VC of 23%, MIP of 15%, and MEP of 16%. These values slightly improved after the initiation of NIV but subsequently showed a declining trend again.

Conclusion

This case highlights the risk of overlooking the neuromuscular cause of respiratory failure in patients after cervical spine surgery. Relying solely on standard pulmonary tests and imaging can lead to missing important indicators, such as decreased supine VC and decreased respiratory muscle strength (MIP/MEP), which are signs of hypercapnia causing respiratory failure. A comprehensive evaluation including supine VC, and respiratory muscle strength measure is essential for early detection. Rapid NIV initiation avoids intubation, improves blood gases, and facilitates recovery even in patients with such severe respiratory failure, allowing patients to maintain their ability to communicate and swallow. This case also highlights the importance of considering underlying neuromuscular diseases such as ALS in patients with persistent respiratory failure.