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Critical Illness-Related Corticosteroid Insufficiency during NIV Training in ALS: A Case Report

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Introduction

Critical illness–related corticosteroid insufficiency (CIRCI) is characterized by inadequate glucocorticoid activity relative to physiologic stress (Figure 1). Although traditionally associated with refractory hypotension in septic shock, recent guidelines emphasize that CIRCI may also present with atypical features, including altered mental status. In pathophysiological perspective, prolonged stress may lead to dysregulation of the hypothalamic–pituitary–adrenal (HPA) axis. In such conditions, cortisol production may become insufficient relative to superimposed physiologic stress, despite preserved adrenal function.

We report a case of acute mental deterioration in an advanced amyotrophic lateral sclerosis (ALS) patient during non-invasive ventilation (NIV) training, in whom relative glucocorticoid insufficiency was clinically suspected and successfully treated empirically.

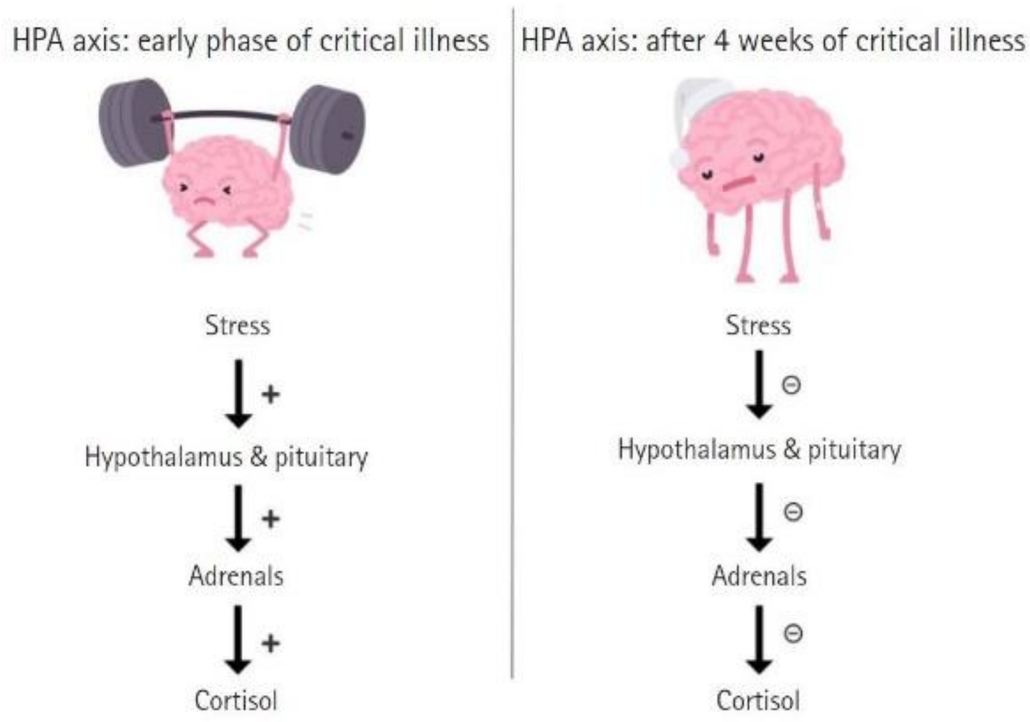

Fig 1. Differential HPA axis responses during early and prolonged critical illness. Adapted from Fredrick FC et al., Acute Crit Care 2024;39:331-340.
* Abbreviation : HPA; hypothalamic-pituitary-adrenal

Case Presentation

A 62-year-old woman with advanced ALS was admitted for respiratory failure. Prior to admission, she had chronically endured respiratory distress at home without respiratory support.

Chest radiography revealed left-sided atelectasis with worsening desaturation and prominent accessory muscle use, requiring intubation and mechanical ventilation in the intensive care unit (ICU). During the ICU stay, marginal blood pressure prompted evaluation for adrenal insufficiency, and demonstrated an adequate cortisol response. However, baseline adrenocorticotropic hormone was at the lower limit of the normal range despite ongoing physiologic stress (Table 1). Because she refused tracheostomy, she was extubated and transitioned to NIV as an alternative strategy.

Parameter	Value	Reference range
ACTH	10.38 pg/mL	10 - 60 pg/mL
Cortisol (basal)	8.61 μg/dL	
Cortisol (30 min)	20.96 μg/dL	≥ 18 μg/dL
Cortisol (60 min)	23.40 μg/dL	≥ 18 μg/dL

Table 1. Rapid adrenocorticotropic hormone (ACTH) stimulation test demonstrating normal adrenal response during ICU stay

Low-dose oral hydrocortisone was administered and continued until hospital day 11. On hospital day 12, during NIV training in general ward, she was found comatose. Vital signs were stable. Arterial blood gas analysis showed normocapnia with adequate oxygenation. Electrolytes and glucose were within normal ranges. Brain imaging revealed no acute lesions, and electroencephalography excluded non-convulsive status epilepticus. Given the clinical context, empirical intravenous hydrocortisone was administered for suspected relative glucocorticoid insufficiency, resulting in improvement within 20 minutes and full recovery within two hours (Figure 2).

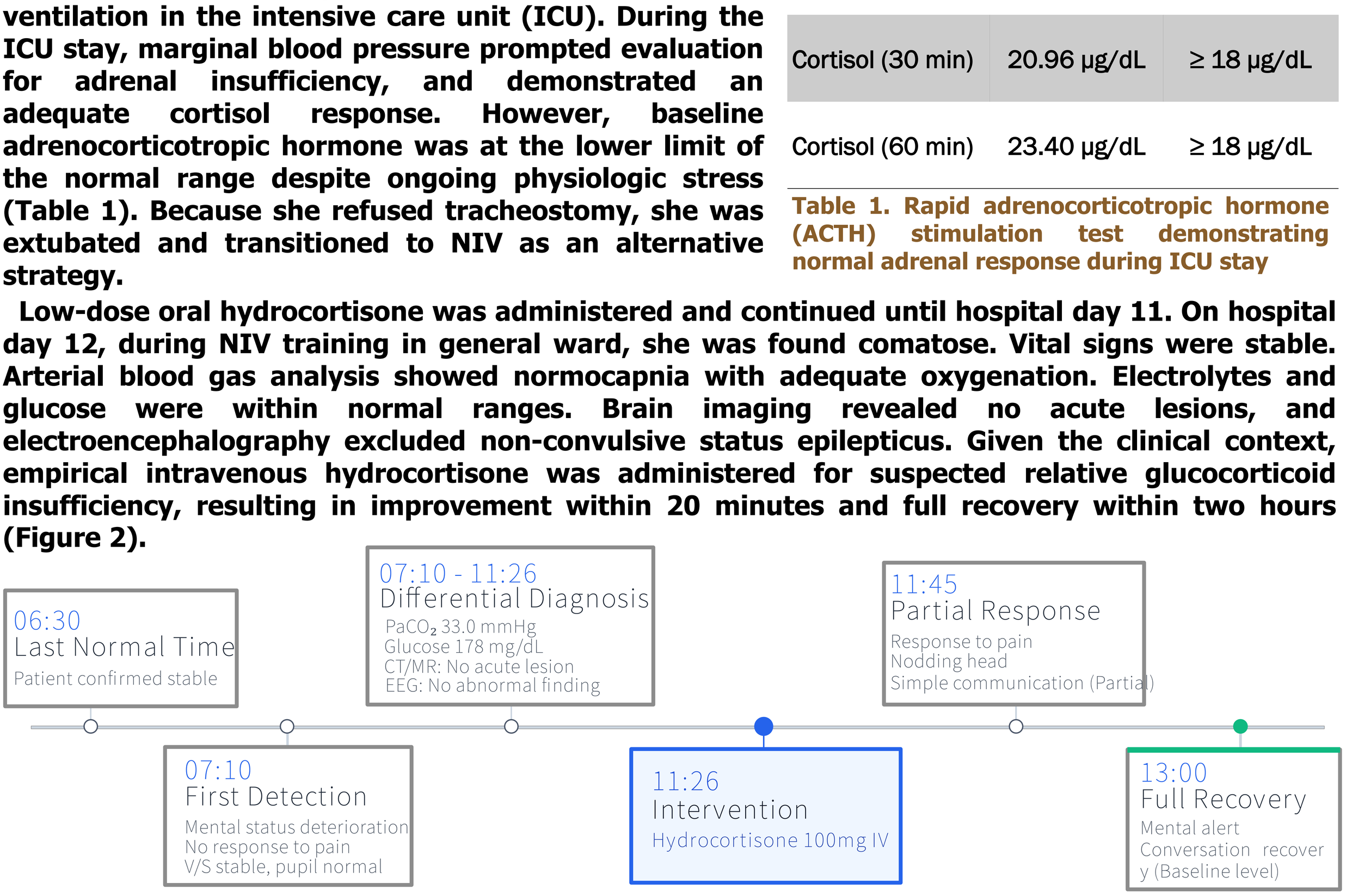

Fig 2. Timeline of acute mental status deterioration and recovery following hydrocortisone administration

Conclusion

NIV training in the general ward requires sustained spontaneous respiratory effort in an alert state and may be accompanied by sleep deprivation and psychological distress, potentially imposing greater physiologic stress than a sedated ICU setting. In advanced ALS patients with chronic progressive respiratory failure, prolonged physiologic stress may contribute to HPA axis dysregulation, whereby superimposed stress during NIV training transiently exceeds neuroendocrine adaptive capacity.

When common causes are excluded, relative glucocorticoid insufficiency should be considered, and empirical corticosteroid therapy may facilitate rapid neurological recovery in selected cases.