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Tetraplegia due to intermittent acute porphyria: the role of physiotherapy in the rehabilitation process – a case report

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Introduction: Acute Intermittent Porphyria (AIP) is one subtype, out of eight, that encompasses the porphyria. Each subtype is secondary to a defect in the pathway of Heme synthesis. AIP is an autosomal-dominant disease, manifested in less than 10% of individuals with the mutation. The clinical presentation varies, and involves a range of symptoms from abdominal pain to important muscular paralysis, with acute depreciation of motor and respiratory functions needing mechanical ventilation and intensive care.^(1,2) In this scenario,

early mobilization and progressive mobility program is an important intervention aiming to improve the functionality and regaining respiratory capacity.^(3,4)

Objective: Report the case of a 38 years old woman, admitted in the Intensive Care Unit (ICU) with decline of respiratory and motor functions, due to the manifestation of AIP. In which, it was possible to optimize the rehabilitation through a progressive mobility program. The study is a case report based on the data obtained through the electronic patient record, evaluations and daily follow-up. It was obtained the Patient Consent for Publication, following the institution regulation.

Case report: Patient feminine, 38 years old with no previous comorbidities and admitted with epigastric pain, progressing in to tetraparesis with braquial and proximal predomination, and respiratory compromise, needing mechanical ventilation. After 5 days intubated, tracheostomy was conducted. The identification of patient's mobility status followed the institution's early mobilization protocol and rehabilitation program was aligned with the medical team. An individualized physiotherapy sessions sheet was developed, prioritizing the weaning ventilation support and motor functionality. The session's plan was reviewed or reformulated weekly based on the periodic evaluation of motor and respiratory functions. During the ICU stay, there was a decrease in the muscle mass, in contrast with progressive improvement of mobility and vital capacity (Figure 1). The weaning of mechanical ventilation and regaining respiratory autonomy was done applying diaphragm electrical stimulation, simultaneously to periods of pressure support ventilation. Evolving to sessions with inspiratory muscle training devices. In the motor rehabilitation branch, the emphasis was neuromuscular electrical stimulation of limbs, tilt table, cycle ergometer and kinesiotherapy progressing from passive, in early stages, progressing to active during ICU stay.

Conclusion: This case report brings to light the importance of individualized and progressive mobility program

in critically ill patients, demonstrating that it may be a key point to obtaining satisfactory improvement in functionality and respiratory capacity.

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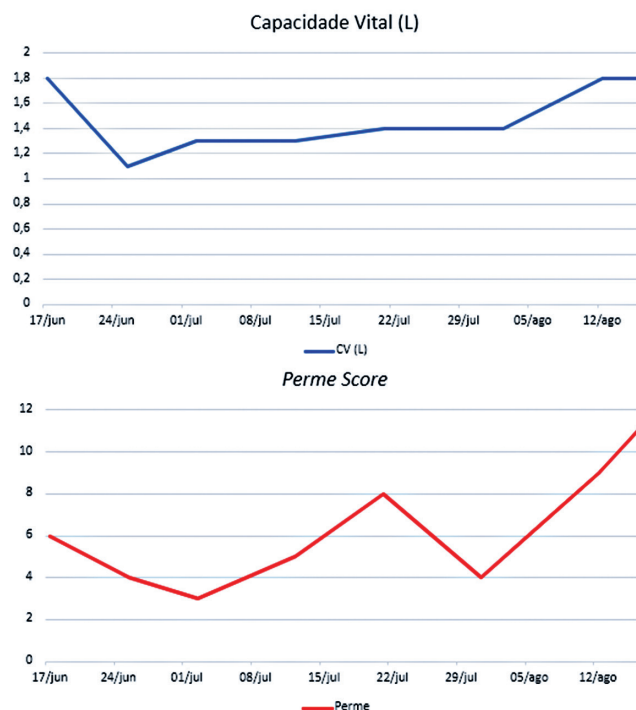


Figure 1. Measures of Vital Capacity (VC) and PERME Score during ICU length of stay