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EDITORIAL

Anaesthesia and Rare Neuromuscular Diseases

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1. INTRODUCTION

Fortunately, early diagnosis, the discovery of new therapies, and the use of a multidisciplinary approach have led to an extension of the life expectancy of patients suffering from rare neuromuscular diseases (NMD). In this group of diseases, both prejudicial and post-judicial pathologies are included [1, 2]. The presence of a neuromuscular pathology is a challenge for the anaesthesiologists and intensivists who have to manage anaesthesia and a perioperative care course in a patient suffering from such characteristic comorbidities. The main focus in the management of the perioperative care of a patient suffering from NMD is the risk of postoperative respiratory failure [3]. These patients could be affected by weakness of the respiratory muscles associated or not with a restrictive respiratory failure due to anatomical alteration of the rib cage. They could also present a poor management of secretions with ineffective cough and/or poor control of the airways, especially in bulbar forms of Amyotrophic Lateral Sclerosis (ALS). It is, therefore, very important to correctly evaluate preoperative respiratory function not only through blood gas analysis but, above all, through the study of vital capacity (VC) and cough peak flow (PCF), the study of sleep apnea-hypopnea with sleep polygraphy possibly related to electroencephalography, as well as maximum inspiratory and exhaling pressure (MIP and MEP) [4]. The evaluation and the management of the airways are also not to be underestimated. These patients could be affected by anatomical alterations (progeny, macroglossia) or by poor control of tongue movement and swallowing. This could lead not only to a problem of ventilation/intubation during general anaesthesia but also to acute respiratory failure due to obstructive effect of the tongue or inhalation during procedural sedation. The latter aspect is easy to be efforted in a center specialized in the treatment of neuromuscular diseases in patients who are required to receive procedural sedation, for example, to perform percutaneous gastrostomies or pacemaker implantation [5].

Another aspect that is very important for the anaesthesiologist and intensivist is the management of tracheostomy. The timing of the procedure is fundamental, as the elective procedure is characterized by a better outcome. It is fundamental to choose the technique (surgical or percutaneous) as well as the ideal anaesthesia for that procedure, with the view that it is possible to perform a surgical tracheostomy by utilizing locoregional anaesthesia. It is important to reduce the hospitalization times in the Intensive Care Unit (ICU) as well as in Postoperative Intensive Care (PACU). These patients are often malnourished, subjected to several previous hospital recoveries, several antibiotic theraies at home, and so are fragile with an enhanced risk of colonization or infection due to the presence of multidrug resistant bacteria. Finally, the need to minimize the risk of procedural tracheal stenosis should be considered. This risk is based on the clinical history of the patient that provides for a periodic replacement of the tracheostomy cannula as well as the high incidence of tracheomalacia [6].

Another interesting aspect of patients suffering from rare neuromuscular diseases is their association with cardiovascular diseases, in particular with rhythm disorders as in the case of Steinert dystrophy, or heart failure as in the case of Duchenne dystrophy and mitochondrial dystrophy. It is, therefore, essential to be prepared for the management of acute arrhythmic complications; also, it is important to not underestimate, in preoperative evaluation of a patient, a simple branch block or type 1 atrioventricular block. In preoperative examinations, the Holter examination may find space in these patients, as many of them may need preoperative pacemaker implantation. Careful preoperative evaluation by the anaesthesiologist is important in case of diseases that are associated with systolic, diastolic or mixed dysfunction, considering the myocardiodepressant and vasodilator effect of anaesthetic drugs (propofol, volatile anaesthetics). Invasive hemodynamic monitoring should thus be employed during the

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induction of anaesthesia. Finally, it is fundamental to not underestimate the risk of malignant hyperthermia or acute rhabdomyolysis in myopathic patients (central core myopathy, calcium channelopathies, King-Denborough syndrome). In addition, succinicolina use not only should be used as a trigger for hyperthermia but also non-dystrophic congenital myotonic forms (Thomsen or Becker) of myotonic seizures associated with difficult ventilation and intubation procedures [7, 8]. Based on these considerations, total intravenous anaesthesia (TIVA) should be the technique of choice. First of all, careful titration of propofol is necessary based on the patient's hemodynamics, and of course, on the bispectral index (BIS) to avoid the administration of a low dosage of sedative. Secondly, the study of the effect of propofol on mitochondrial neuromuscular diseases is interesting since the OXPHOS complex I inhibitor effect and/or the inhibitory beta-oxidation effect could be the basis of propofol infusion syndrome (PRIS). This syndrome is characterized by metabolic acidosis, severe heart failure, fever and muscle cell lysis. Finally, a comment must be made on the role of locoregional anaesthesia. While there are reservations in the literature as to whether the peripheral nervous system is exposed to possible damage from needles, catheters, vasopressors, or direct toxicity from local anaesthetic, there is evidence that locoregional technique reduces postoperative complications related to dysventilation and that the routine use of ultrasounds reduces the potential risk of damage to nerve fibre [9].

Another very interesting aspect for intensivists who approach an NMD patient is the knowledge of the patient's nutritional needs and the metabolic specificity of these patients characterized by different states of disease and progressive immobilization. Harris Benedict's formula must be adopted with regard to the state of mobilization and physical activity and hours of mechanical ventilation. Theoretically, a specific enteral formula should be characterized by high caloric content and low carbohydrate content to avoid the excessive production of carbon dioxide. In this case, indirect calorimetry may be utilized [10].

CONCLUSION

Finally, it is very difficult to interpret with the usual parameters the kidney function, as all neuromuscular patients with depletion of the muscles present themselves with a very low level of circulating creatinine; thus, the evaluation of the volemic status with the help of echocardiography or invasive monitoring systems and the careful examination of the amount of urine produced and the dosage of cystatin C are fundamental. Based on all these considerations, the presence of multidisciplinary reference centers for the treatment of neuromuscular pathologies is thus important, and the anaesthesiologist and the intensivist constitute an integral part of the team of care for patients suffering from this rare and extremely complex pathology.

CONFLICT OF INTEREST

Dr. Vincenzo Pota is the Associate Editorial Board Member for The Open Anesthesia Journal.

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