Introduction

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Muscle diseases are relatively uncommon, but patients presents to operating room for diagnostic studies, for treatment of complications, or for surgical management of unrelated disorder. Although the causes of these diverse diseases are frequently unknown, it has been possible in most cases to place the defect at or beyond the neuromuscular junction.

Patients with muscle disease who are capable of fairly normal activities may have decreased muscle reserves to compensate for the stress of anesthesia and operation. Thus, they are analogous to a partially "curarized" patient who seems normal but for whom any added stress may be catastrophic ***(Benumof, 1998)***.

The past several years have brought an explosion in knowledge concerning the molecular basis of muscular disorders. Not only has information about the specific genetic defects been bountiful, but also new insights into the pathomechainsms have been gained ***(Nagiub et al.2002)***.

Because of fear of postoperative respiratory failure in these patients, a preoperative base line measurement of respiratory reserve with at least simple tests of lung function (such as vital capacity & maximum inspiratory and expiratory force) and blood gas analysis will allow better postoperative assessment of these parameters. The use of pulse oximeter that is readily available non-invasive tool frequently reveals abnormal lung function before surgery while the patient is still breathing room air.

The measurement of serum enzymes derived from muscle is most abnormal in patients with ongoing muscle degeneration. However, enzymes are frequently normal in severe muscle diseases that don't have continuous muscle destruction.

Preoperative use & teaching of chest physiotherapy may avoid many problems in the postoperative period which can be life saving.

The anesthetic requirements are similar to these for any patient, but the margin of safety is reduced. One should use the smallest amount of agents that provide satisfactory operating conditions. Fixed dosing schedules should be avoided, and continuous adjustments should be made according to the demands of the situation ***(Benumof, 1998)***.

It should be remembered that narcotics are as much a potential source of respiratory difficulty, as are general anesthesia and muscle relaxants.

Although succinylcholine is a valuable and preferred relaxant in certain situations, its use presents small but significant risk, as it may cause an abrupt cardiac arrest in patients with occult muscle disease ***(Frank, 1995)***.

The patient must demonstrate adequate respiratory reserve and an ability to handle secretions before leaving the recovery room. If either of these criteria is not met, mechanical ventilation via an endotracheal tube should be considered.

Occasionally, patients with oropharyngeal weakness and are incapable of taking care of there own secretions and maintaining a patent airway will maintain adequate vital capacity and blood gas parameters

with an endotracheal tube in place only to suffer rapid decompansation on extubation. If prolonged ventilalatory assistance is required, tracheostomy may be necessary.

Respiratory support using nasal continuous positive airway pressure (CPAP) may avoid tracheal intubation in the postoperative period.

Early extubation, if possible, seems warranted by the published case reports, though it must be understood that the patient may tire and require reintupation after many hours of apparently normal ventilation. The patient could be given a trial of spontaneous ventilation when the patient has an inspiratory effort greater than –30cm H_2O and a vital capacity of at least 15ml/kg body weight. The patients is then allowed to maintain ventilation for 1 to 2 hours, during which time arterial blood gases are checked to ensure that there is no fatigue. In addition vital capacity measurement and clinical assessment continue. Under these conditions, anticholinesterase therapy may be reinstituted and the patient may be extubated. Even then, it is mandatory that respiratory support be available immediately.

In the postoperative period, Careful monitoring of both objective & subjective parameters must continue. Those patients should be managed at intensive care unit as they may be particularly difficult to wean.

Although most anesthesiologists have only limited experience with muscle disease, this essay will try to enlighten some facts about such diseases & their anesthetic management.