Preparedness of Nurses for Malignant Hyperthermia

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Preparedness of Nurses for Malignant Hyperthermia
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Introduction

Malignant hyperthermia (MH) is a potentially lethal genetic disorder that occurs following exposure to certain triggering agents. The signs and symptoms of MH are associated with the skeletal muscle that causes a shift in intracellular calcium. The people who are susceptible to MH have a gene mutation in the ryanodine receptor subtype 1 (RYR-1) gene which is found in all skeletal muscle. This gene encodes the calcium ion channel in skeletal muscle and is responsible for calcium entry into the myoplasm. The RYR-1 is also the binding site for inhaled anesthetics and the receptor for several muscle relaxants.

Pathophysiology

Malignant hyperthermia is a rare, autosomal dominant genetic condition that occurs within minutes or up to an hour after the administration of the triggering agent. The signs and symptoms of MH are associated with an increase in intracellular calcium. This increase leads to rapid cell death and release of potassium into the bloodstream.

Signs and Symptoms

• Hypercarbia
• Cardiac dysrhythmias
• Hypoxia
• Tachycardia
• Hyperthermia
• Flushed skin
• Cyanosis
• Hyperkalemia
• Generalized muscle rigidity
• Hypotension
• Dyspnea
• Coma

The signs and symptoms of malignant hyperthermia usually occur within minutes or up to an hour after the administration of the triggering agent. However, if a triggering agent is used in conjunction with an inhalational anesthetic, the onset of symptoms is generally within 5 minutes of the administration of the triggering agent. The rapid increase in body temperature can exceed 43.7°C (110.7°F) and can increase by 1°C to 2°C (1°F to 3°F) every five minutes. Malignant hyperthermia cases usually occur as a result of the administration of dantrolene. The contracted cells eventually deplete the oxygen and ATP and begin to produce energy anaerobically. The rapid increase in body temperature can exceed 43.7°C (110.7°F) and can increase by 1°C to 2°C (1°F to 3°F) every five minutes.

The goal of treatment for malignant hyperthermia is to reverse the hypermetabolic state and prevent the potentially lethal consequences of MH. The literature indicates that after diagnosis of MH with appropriate treatment, mortality is very low. The most effective treatment for MH is the administration of dantrolene sodium. The mortality rate from MH used to be as high as 80%, but since the discovery of the MH1 gene, it is now down to 5% (Hirshey Dirksen et al., 2013, p. 330). Dantrolene is a specific ryanodine receptor antagonist and reverses the MH-related muscle contractions by decreasing the calcium in muscle cells (Seifert et al., 2014, p. 192). The longer time that elapses between the onset of MH and the first dose of dantrolene results in an increased risk of complications associated with MH. It is well known that the preparation of dantrolene is difficult. Some of the difficulties with dantrolene preparation are that it requires large quantities of diluents to be mixed and a total of 125 mg of dantrolene per milliliter of diluent.

Significance of Pathophysiology

The significance of the MH1 gene is that it is the causative genetic factor associated with malignant hyperthermia. The MH1 gene is a mutation in the ryanodine receptor subtype 1 gene (RYR-1). The RYR-1 gene is responsible for calcium release within the myocyte. The RYR-1 is also the binding site for inhaled anesthetics and the receptor for several muscle relaxants. The MH1 gene mutation in the ryanodine receptor subtype 1 gene (RYR-1) is responsible for calcium release within the myocyte.

Conclusions

Malignant hyperthermia is a very rare condition that occurs approximately once in every 5,000 to 5,500 anesthetics where volatile anesthetics are administered (Bircher et al., 2013, p. 130). It is of importance, though, because it is one of the few life-threatening situations and is a medical emergency. It is unique in that it is the only condition in which many nursemare responsible for caring for a patient with MH. The purpose of this manuscript is to discuss the signs and symptoms, diagnosis, treatment, and location of MH. The MH1 gene mutation may be monitored by personnel who may be affected. The signs and symptoms may include hypoxia, tachycardia, sinus tachycardia, and ventricular fibrillation (Stewart, 2014, p. 253). Other possible signs and symptoms of MH may include hyperthermia, tachycardia, hypotension, dyspnea, hyperkalemia, and pulmonary edema, rales, and frothy sputum.

References


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