Malignant Hyperthermia

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Malignant hyperthermia
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Definition

Malignant hyperthermia (MH) is an autosomal dominant disorder that is inherited. The disturbance of calcium homeostasis associated with MH affects skeletal muscle (Schneiderbanger, Johannsen, Rowser, & Schuster, 2014). This hypertrophic muscular disorder is triggered by exposure to depolarizing muscle relaxants such as succinylcholine or halogenated volatile anesthetic gases (Koch, 2012; King & Denborough, 2013). When the dysregulated response is triggered to exposure to a triggering agent, such as halogenated volatile anesthetics and succinylcholine, a depolarizing neuromuscular blocking agent, a core disorder of MH occurs, resulting in the uncontrolled release of calcium within the cell (Schneiderbanger et al., 2014). This releases calcium from the sarcoplasmic reticulum, leading to a hypermetabolic state in the susceptible individual (Kim, 2012).

Underlying Pathophysiology

Individuals with MH have a mutation of the ryanodine receptor subtype 1 (RYR1) gene on chromosome 19p13.1-13.2 (Li, Brantly, & Seibert, 2011; Thomas & Crowhurst, 2013). When the dysregulated receptor is exposed to triggering agents, such as halogenated volatile anesthetics and succinylcholine, a depolarizing neuromuscular blocking agent, a core disorder of MH occurs, resulting in the uncontrolled release of calcium within the cell (Schneiderbanger et al., 2014). This results in the uncontrolled release of calcium and results in a hypermetabolic state in the susceptible individual (Kim, 2012).

Underlying Pathophysiology

As a result of this uncontrolled release of calcium and continuous muscle contraction, muscle rigidity occurs. As these events continue, cellular adenine triphosphate is depleted resulting in posttetanic muscle rigidity and eventually rhabdomyolysis. Rhabdomyolysis occurs when cell contents such as creatine phosphokinase, potassium, and myoglobin are released into circulation due to the deterioration of the cell membrane. Also, in a MH event, oxygen consumption is increased due to the continuous activation of aerobic and anaerobic metabolism. This increase in oxygen consumption results in acidosis, hyperglycemia, increased body temperature, and excessive production of CO₂ (Schneiderbanger et al., 2014). Figures 1 and 2 illustrate the pathophysiological changes that occur during a malignant hyperthermia event.

Signs & Symptoms

Table 1 Clinical signs of malignant hyperthermia (Schneiderbanger et al., 2014, p.350)

<table>
<thead>
<tr>
<th>Early Signs</th>
<th>Late Signs</th>
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<tbody>
<tr>
<td>Masseter spasm</td>
<td>Hyperthermia</td>
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<tr>
<td>Generalized muscular rigidity</td>
<td>Rhodomyolysis</td>
</tr>
<tr>
<td>Tachycardia (&gt;80%)</td>
<td>Acute renal failure</td>
</tr>
<tr>
<td>Hypercapnia</td>
<td>Cardiac arrhythmia</td>
</tr>
<tr>
<td>Hypoxemia</td>
<td>Hypotension</td>
</tr>
<tr>
<td>Combined metabolic-respiratory acidosis</td>
<td>Circulatory failure</td>
</tr>
</tbody>
</table>

Implications for Nursing Care

- MH treatment objectives: stop the abnormally metabolite reaction with dantrolene and restore normal hemodynamic conditions, temperatures, and metabolic balance (Seifert, Wahr, Pace, Cochrane, & Scholtis, 2014).
- Recommended management approach: Utilize a multidisciplinary medical team trained to promptly and accurately diagnose MH, work cooperatively, understand multiple roles and responsibilities, and efficiently utilize readily available equipment and medications; medical teams should periodically review MH protocols and participate in mock drills to practice tasks and roles (Dirksen et al., 2013).
- See Figure 2 for recommended roles and treatment.

Crisis Considerations: Malignant Hyperthermia

The ultimate goal is to prevent the occurrence of MH. As discussed by Beggs, McCann, and Powers (2012), prior to the administration of anesthesia, obtaining thorough patient and family histories related to adverse anesthesia reactions may alert the anesthesiologist to the patient’s possible predisposition to MH and dictate the selection of non-triggerring MH anesthetics. However, when prevention is not realized, continued vigilance and crisis preparedness on the part of multidisciplinary medical team members will further reduce the MH mortality rate and improve patient outcomes (Schneiderbanger et al., 2014).

References