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Malignant Hyperthermia: The Perioperative Nurse's Role

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ABSTRACT

The malignant hyperthermia (MH), a rare but life-threatening condition triggered by certain anaesthetic agents in patients with a genetic predisposition. MH is characterised by a hypermetabolic response, leading to symptoms including increased carbon dioxide levels, muscle rigidity, irregular heart rhythms, and hyperthermia. Epidemiological data indicate a higher prevalence in children, males, and specific populations such as those of French, Scandinavian, and Japanese origin. The diagnosis of MH is challenging and the gold standard is the halothane-caffeine muscle contracture test, though genetic testing is also utilised. However, not all associated gene mutations are fully identified and testing can be costly and invasive. Risk factors include age, genetics, type of anesthetic agents, and environmental triggers. The symptoms manifest in two distinct phases: the early phase, characterised by elevated end-tidal carbon dioxide levels, muscle spasms, and metabolic acidosis; and the late phase, marked by hyperthermia, rhabdomyolysis, and cardiac arrhythmias. Effective management of MH necessitates two fundamental components: the early recognition of symptoms and the prompt response of a well-coordinated perioperative team. The immediate measures to be taken include the discontinuation of triggering agents, the administration of 100% oxygen, the initiation of active cooling measures, and the intravenous administration of dantrolene sodium to control the crisis. The monitoring of vital signs, electrolyte balance, and kidney function is crucial. Post-crisis care involves prolonged observation to prevent recurrence, rehabilitation, and patient education. The role of perioperative nurses is pivotal in identifying early signs, initiating rapid intervention, and providing ongoing care. Institutions must maintain protocols, ensure the availability of MH treatment trolleys, and provide regular training to staff. Risk assessments and identification of susceptible patients through family history or genetic testing are critical preventive measures. Early detection, prompt treatment, and coordinated care are essential to improving patient outcomes in MH cases.

Keywords: Malignant hyperthermia, nursing care, surgical nursing.

INTRODUCTION

Malignant hyperthermia (MH) is a rare but life-threatening complication which develops as a hypermetabolic response to specific anaesthetic agents used during anaesthesia (halothane, isoflurane, sevoflurane, desflurane) and succinylcholine, which is a muscle relaxant and is related to the genetic factors of the patient (1-5). The first documented instance

of malignant hyperthermia was by Gudel in 1937, and 1960, Denborough and Lovell reported that hereditary factors were effective in its development (6). The Malignant Hyperthermia Association of the United States (MHAUS) was established in 1981 due to an increase in the incidence of malignant hyperthermia (7). The North American Malignant Hyperthermia Registry (NAMHR) was established in 1987 and merged with MHAUS in 1995, ensuring comprehensive patient data collection and facilitating research in this area (8). The website prepared by MHAUS provides healthcare professionals with resources on a range of subjects, with a particular focus on crisis management procedures. Simultaneously, patients are informed and recorded (6). Although malignant hyperthermia is a rare complication in terms of patient safety, perioperative team members must perform crisis management most optimally. The primary objective is to eliminate the threat to the patient's life and to ensure the continuity of emergency treatment and care. Nurses, a pivotal component of the perioperative team, assume significant roles and responsibilities in this regard. Therefore, nurses must possess a comprehensive understanding of the epidemiology, risk factors, diagnostic criteria, signs and symptoms, treatment, and care process of MH.

Epidemiology

The incidence of MH in adults has been documented to range from 1/10,000 to 1/100,000 to 250,000 (7,9). According to data from the Royal College of Anaesthetists National Audit Project, the prevalence of MH in the UK has been reported as 1/100,000 (5,10,11). Furthermore, a gender disparity has been observed, with the prevalence of MH reactions being two times higher in males than in females (12,13). While MH affects all races, it has been reported that MH susceptibility is significantly higher in French, Scandinavian and Japanese populations (3). Concerning age, although MH is observed more frequently in the paediatric population than in adults, MH can be observed in one out of every 30,000 surgeries in children (14,15). The ability to predict a patient's predisposition for developing MH complications is limited; it is impossible to determine this before many patients are exposed to the relevant anaesthetic agent and experience an MH crisis. It has been documented that in some cases, MH may occur following at least three exposures to the triggering agent (6,14,16).It is recognised that there are more than 80 genetic disorders associated with MH in individuals who are sensitive to anaesthetic agents, leading to the development of MH (7).Although many of the gene mutations have been characterised, there are still different mutations that have not been defined. Furthermore, it has been observed that some patients who are genetically predisposed to MH do not develop the condition despite exposure to the triggering agent (15,17).

Risk factors

There are numerous documented risk factors that can lead to malignant hyperthermia. However, research has indicated that the type of anaesthetic agent, age, genetics, family history, environmental temperature and stress are frequently effective (6,9). Exercise has been documented as a trigger for this condition, albeit infrequently (4). Furthermore, soft tissue infection and muscle injury have been documented as risk factors for MH crisis (6).

Signs and Symptoms

An unexplained and unexpected increase in end-tidal carbon dioxide (etCO2), increased heart rate and increased temperature after anaesthesia. In MH, the clinical picture starts suddenly and progresses very rapidly. The first symptom is an unexplained increase in carbon dioxide. At the same time, the oxygen requirement of the body increases 2-3 times with excessive acceleration of metabolism. The body consumes excessive amounts of oxygen and

glucose, and produces carbon dioxide and heat (9).Cardiac arrest and death may occur as a result of tension in the heart muscles with rapid oxygen consumption (6). In susceptible patients, masseter muscle spasm occurs rapidly in the first 60-90 seconds after inhalation of triggering inhaler anaesthetic (5). Abnormal and uncontrolled release of calcium ions from the sarcoplasmic reticulum in the skeletal muscle results in continuous and involuntary contractions, causing cell damage and rhabdomyolysis. This, in turn, leads to disruption of the heart's rhythm, with the release of high amounts of potassium into the blood. Additionally, myoglobin resulting from muscle destruction can damage the kidneys.

In MH, fever manifests as a late symptom. The body temperature increases by one degree every five minutes, reaching a maximum of 44^0C (6). The manifestation of symptoms in MH occurs in two distinct periods: the early and late phases. A comprehensive overview of symptoms and findings can be found in Table 1 (5,7,18,19).

Table 1. Signs and Symptoms of Malignant Hyperthermia

Early Period	Late Period
The following symptoms were observed: -Elevated levels of endtidal carbon dioxide -Increased oxygen consumption -Elevated heart rate -Increased respiratory rate -Irregular arterial blood pressure -Masseter muscle spasms -Diffuse muscle rigidity -Rhabdomyolysis -Increased sweating -Metabolic acidosis -Respiratory acidosis -Skin mottling -Arrhythmia	-Hyperkalaemia -Rapid increase in body temperature -Excessive elevation of creatinine kinase levels -Excessive elevation of myoglobin levels -Darkening of urine colour due to myoglobinuria (black urine) -Serious cardiac arrhythmias -Disseminated intravascular coagulation (DIC) -Cardiac arrest -Acute renal failure

Diagnostic Methods

The gold standard for the diagnosis of MH is the halothane-caffeine-induced in vitro contracture test with muscle biopsy (4,9). In this test, the patient's muscle tissue is exposed to halothane-caffeine and the resulting muscle tension is measured. A positive response to the halothane-caffeine test is indicative of susceptibility to MH, whereas a negative response rules out this possibility. However, it should be noted that the test is considered invasive and costly, which limits its widespread use (17,19). Malignant hyperthermia is an inherited condition that is carried by an autosomal dominant gene. Family members of an individual with malignant hyperthermia must be also investigated.Genetic testing is available in select centres across the United States of America (7). Notwithstanding, given that not all gene mutations associated with MH have been fully defined, negative results from genetic tests do not exclude the possibility of malignant hyperthermia in an individual patient (6).

Some patients have a high risk of developing MH. Patients with a high risk of developing MH with exposure to a triggering agent include those with the following characteristics:

- A presence of pathological genetic mutations
- A positive in vitro muscle contracture test
- A personal and family history that may affect MH
- Patients with myopathy of genetic aetiology causing MH sensitisation
- An unknown variant in genes causing MH susceptibility
- A history of recurrent rhabdomyolysis

- People with unexplained exercise-induced heat disease (5,20-22).

The Malignant Hyperthermia Clinical Grading Scale (MHCGS) was developed by Larach (1994) to evaluate malignant hyperthermia. It is widely accepted as a standard clinical tool, incorporating six dimensions: muscle stiffness, muscle breakdown (as indicated by an increase in creatine kinase level), respiratory acidosis, temperature increase, cardiac involvement and family history. The scale's total score determines the likelihood of MH, with probabilities ranging from rarely to almost certain. The evaluation of MH by nurses is facilitated by calculating the scale score (23, 24).

Treatment and Nursing Care

Although malignant hyperthermia is a rare complication, it may result in death if not controlled in the early period. Early diagnosis, emergency treatment and care are important in the prevention of complications (2,7). The principles of treatment and care that should be applied in case of MH are as follows:

- It is imperative that effective cooperation between the perioperative team is ensured.
- The following predefined roles must be fulfilled by perioperative team members: team leader, medication nurse, registrar, laboratory liaison, intravenous access nurse, lavage nurse and staff roles such as ice liaison. In the event of MH reaction, the anaesthesiologist, surgeon and other perioperative team members must be informed immediately and assistance requested.
- Discontinuation of all triggering agents is imperative.The vaporiser must be switched off and removed from the anaesthesia machine.
- It is imperative that no time is lost in changing the anaesthesia machine. The anaesthesia machine should be flushed using >10 litres/min of oxygen and air for 90 seconds. Activated carbon filters should be placed on the inspiratory and expiratory parts of the anaesthesia machine to absorb volatile anaesthetic substances, and the breathing circuit and soda-lime box should be replaced.
- Furthermore, it is imperative to provide 100% oxygen at maximum flow.
- Hyperventilation should be facilitated by targeting normocapnia (minute ventilation should be increased 2-3 times the normal value), and the etCO2 level should be monitored.
- The MH trolley, equipped with standard MH equipment and the MH treatment protocol, should be promptly transferred to the patient.
- Preparations for the placement of arterial and central venous catheters should be initiated immediately, and intravenous access should be established using wide cannulas. The arterial catheter should then be inserted.
- Intravenous administration of dantrolene sodium is also recommended.
- Monitoring of central body temperature is imperative.
- Furthermore, the use of intravenous fluid warmers and hot air blower heaters should be discontinued.
- Active body cooling should be initiated, encompassing the administration of cold intravenous fluids, the application of cooling blankets, the insertion of three-way urethral catheters, and the support of active cooling by continuous bladder irrigation or lavage with iced saline through nasogastric tubes. Additionally, the room air conditioner should be activated. Bilateral application of ice packs to the groin and armpits is also recommended.
- If deemed appropriate, surgery should be terminated, postponed or anaesthesia should be continued with safe drugs (regional anaesthesia, local anaesthesia, total intravenous anaesthesia, non-depolarising muscle relaxants, opioids, sedatives)
- The patient should be monitored and vital signs should be monitored.

- The insertion of a central venous catheter should be considered for the assessment of fluid volume and central venous pressure. Monitoring of electrolyte balance is also recommended.
- The implementation of symptomatic treatment is imperative.
- In the event of cardiac arrest, the advanced life support algorithm should be followed and the patient should be closely monitored for 48-72 hours as MH may recur in 25% of patients following return of spontaneous circulation
- Arterial blood gas and other laboratory findings (potassium, calcium, creatine kinase, myoglobin, glucose, coagulation factors) should be monitored.
- Severe acidosis should be corrected, and hyperkalaemia should be treated.
- In patients exhibiting masseter muscle spasms, signs and symptoms of rhabdomyolysis should be monitored for a period of 12-24 hours. At 24 hours, a creatine kinase test should be performed. Monitoring of myoglobinuria and urine colour is to be achieved by means of a urinary catheter. Furthermore, the presence of acute renal failure and compartment syndrome must be closely monitored.
- In cases of suspected malignant hyperthermia, consultation with a specialist centre should be considered for further advice and ongoing monitoring.
- It is imperative to exercise caution and monitor for potential treatment-related adverse effects.
- In the aftermath of the attack, it is imperative to closely monitor patients for signs of muscle weakness and elevated pain levels, a consequence of muscle destruction and the administration of dantrolene sodium. The implementation of necessary treatments and the initiation of a comprehensive rehabilitation programme are crucial. It is essential to inform the patient and their relatives that the restoration of pre-attack muscle strength may require a protracted period, spanning weeks to months (2,3,5-7,25-31).

CONCLUSION

Malignant hyperthermia is a rare condition, but one which can prove life-threatening. The following measures are to be implemented for the purpose of prevention, treatment and care in the perioperative period:

Firstly, it is imperative to recognise the MH crisis as an emergency that demands effective team coordination. Team members must be thoroughly versed in the institution's established protocols and their designated roles. Organising in-service training and ensuring personnel are kept up to date with the latest knowledge are essential components of this process. The institution should create and rehearse protocols with team members, and any missing areas should be addressed.

The MH trolley, which contains the materials to be used during intervention in the MH crisis and a sufficient supply of dantrolene sodium, should be maintained in the hospital. The MH trolley should be subject to regular checks. Furthermore, it is essential to ensure the availability of all necessary fixtures and consumables for cooling and follow-up procedures.

It is imperative that patients deemed to be at risk of MH are identified and that their information is meticulously documented by an association or centre. Furthermore, it is imperative that the families of these patients undergo screening tests.

Risk assessment for malignant hyperthermia should be performed using scoring systems in the perioperative period. In instances where surgical intervention is deemed necessary for a patient with a high risk of MH, they must be monitored closely within the hospital's early warning system. Individuals deemed to be at risk should be identified through the use of wristbands or necklaces.

Operating theatre nurses must observe patients during surgery and be aware of the early signs of MH. The recognition of symptoms by operating theatre nurses should be followed by the alerting of the entire surgical team and the initiation of rapid intervention.

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