

## Malignant hyperthermia in a 6-month-old infant

### ABSTRACT

Malignant hyperthermia (MH) is a rare hypermetabolic disorder of skeletal muscles that manifests as a life-threatening crisis in susceptible individuals, after exposure to triggering agents, most commonly halothane and succinylcholine. MH presents with multiple nonspecific signs and laboratory findings such as tachycardia, hyperthermia, hypercarbia, acidosis, and muscle rigidity. Caffeine halothane contracture test is not available at most centers in India. Larach *et al.* have described a clinical grading scale for determining the MH raw score based on clinical findings and biochemical tests. The high degree of suspicion, early recognition and aggressive treatment should commence immediately. It is imperative to avoid triggering agents, such as volatile anesthetics and succinylcholine, and promote the use of total intravenous anesthesia in MH susceptible patients. We report a case of 6-month-old child undergoing laparotomy under general anesthesia, who presented with signs and symptoms of MH, had MH rank 5 and raw score 36.

**Key words:** Halothane; malignant hyperthermia; malignant hyperthermia clinical grading scale; succinylcholine

### Introduction

Malignant hyperthermia (MH) is a hypermetabolic disorder of skeletal muscles that manifests as a life-threatening crisis in susceptible individuals, after exposure to triggering agents, most commonly halothane and succinylcholine.<sup>[1,2]</sup> Incidence in pediatric patients is 1 in 15,000 anesthetics, but it is rare in infants.<sup>[3]</sup> MH presents with multiple nonspecific signs and laboratory findings such as tachycardia, tachypnea, hypercarbia, respiratory and metabolic acidosis, and masseter muscle rigidity, the rapid increase in body temperature, hyperkalemia, and hemodynamic instability. Therefore, MH clinical grading scale proposed by Larach *et al.* is used to rank the likelihood that an adverse anesthetic event may represent MH.<sup>[4]</sup> Here, we present a case of 6-month-old infant showing signs and symptoms of MH after exposure to halothane and succinylcholine.

### Case Report

A 6-month-old female child (7.2 kg) was scheduled for emergency laparotomy for intussusception. Her birth and developmental history were unremarkable, and there was no previous history of surgery or anesthetic exposure. There was no family history of anesthetic complication or neuromuscular disorder. Preoperative pulse rate (PR) was 120/min, noninvasive blood pressure (NIBP) 86/56 mmHg, oxygen saturation (SpO<sub>2</sub>) on room air 99%, and skin temperature 99°F. The patient was premedicated with injection glycopyrrolate 0.075 mg and tramadol 15 mg intravenously through 24G intravenous (IV) cannula and isolyte P was started. After preoxygenation for 3 min, injection propofol 15 mg followed by succinylcholine 15 mg IV were given to facilitate intubation. No generalized muscle rigidity or masseter spasm was noted, and the

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patient was intubated with 3.5 mm uncuffed endotracheal tube using rapid sequence induction without any difficulty. After induction, all vital parameters including NIBP, PR, SpO<sub>2</sub>, electrocardiogram, end-tidal carbon dioxide (EtCO<sub>2</sub>), and temperature were within normal limits. Anesthesia was maintained with halothane 0.6-1% in 100% oxygen and injection atracurium (loading dose 0.5 mg/kg followed by maintenance dose of 0.1 mg/kg). Half an hour later, EtCO<sub>2</sub> started increasing up to 45-50 mmHg. In order to bring EtCO<sub>2</sub> back to normal, fresh gas flow and minute ventilation were increased and soda lime was replaced, but the EtCO<sub>2</sub> continued to rise. Suspecting MH crises halothane was stopped, and its vaporizer was removed. Anesthesia was maintained with fentanyl 2 µg/kg/h and propofol 2 mg/kg/h infusion. The Patient had normal temperature at this time, but PR rose to 150/min. Within next half an hour, EtCO<sub>2</sub> rose to 60 mmHg, thereafter in next 15 min up to 75 mmHg and patient developed sinus tachycardia with the heart rate of 180/min and nasopharyngeal temperature of 103°F. As per European MH group guidelines, measures to decrease the temperature were carried out including IV cold saline drip, cold sponging, and gastric lavage with cold saline. IV paracetamol loading dose 20 mg/kg followed by 5 mg/kg infusion was given. Meanwhile, blood sample for serum electrolytes and arterial blood gas were sent and surgeon alerted regarding the possibility of MH. Gradually, temperature decreased to 100°F, PR to 140/min EtCO<sub>2</sub> to 60 mmHg in the duration of 30 next min. Surgery was completed, and all anesthetic agents were stopped. Neuromuscular blockade was reversed with neostigmine 0.07 mg/kg and glycopyrrolate. Suddenly, the patient developed seizures which were controlled with thiopentone sodium 15 mg IV. Arterial blood gas revealed metabolic acidosis (pH -7.2), hypercarbia (PaCO<sub>2</sub> -84 mmHg), and hyperkalemia (K+ 7.0 mEq/l). Calcium chloride 0.1 mmol/kg slow IV was given to correct hyperkalemia and injection furosemide 1 mg/kg was given to prevent renal injury. The patient was continuously hyperventilated for the correction for respiratory acidosis. The patient was extubated when muscle power was adequate, SpO<sub>2</sub> 98% and EtCO<sub>2</sub> of 35-42 mmHg. She was shifted to Intensive Care Unit, where she was oxygenated and monitored for next 24 h.

The patient was kept under observation in the ward during that she remained normal and then discharged on the 7<sup>th</sup> postoperative day. Her attendants were cautioned about possible danger of MH in subsequent anesthesia and she was referred to higher center to further investigate the cause of this hyperthermia episode, as muscle biopsy for MH and urinary vanillylmandelic acid for pheochromocytoma are not available at our institute.

## Discussion

Due to the presence of raised EtCO<sub>2</sub>, hyperthermia and sinus tachycardia differential diagnosis of thyroid storm, pheochromocytoma, and neuroleptic malignant syndrome was considered. No signs of thyrotoxicosis or enlarged thyroid in either mother or child on preoperative examination ruled out thyroid storm. Pheochromocytoma was not likely as blood pressure was normal, and there was no relevant abnormality on abdominal ultrasonography. Neuroleptic malignant syndrome was ruled out as the patient was not on any medication that are implicated for this condition. Thus, a provisional diagnosis of MH was made according to Larach clinical grading scale for MH. This scale ranks the likelihood that an adverse anesthetic event represents MH. Clinical and laboratory findings are assigned points, which are summed up to give a raw score, which is then translated to MH rank. MH rank indicates the likelihood of MH from 1 (almost never) to 6 (almost certain). Our patient had a raw score of 36, which put her in MH rank 5, making the diagnosis of MH very likely [Table 1].

Plasma creatine kinase and myoglobin levels could not measure due to unavailability of these tests during nonroutine hours at our center.

## Conclusion

To conclude, in spite of high clinical variability of MH specially in young children, outcome can be improved by using mandatory monitoring such as temperature probes, EtCO<sub>2</sub> and using MH crisis management checklist.<sup>[6-8]</sup> Thus, the high degree of suspicion, early recognition, and prompt treatment are instrumental in saving the patient's life in spite of nonavailability of dantrolene. Furthermore, it is imperative to avoid triggering agents, such as volatile anesthetics and succinylcholine, and use total IV anesthesia in suspected cases so as to reduce or limit the degree of MH reaction. Specialized tests for the diagnosis of MH such as *in vitro* contracture testing (IVCT) have inconsistent results in young children and infants.<sup>[5]</sup> As a surrogate, the parents are tested by IVCT and genetic diagnostic. Since these tests are available

**Table 1: Clinical indicators for calculating MH raw score in our patient**

Process	Indicator	Points
Rigidity	None	0
Muscle breakdown	Serum potassium >6 mEq/l	3
Respiratory acidosis	PETCO <sub>2</sub> >55 mmHg with controlled ventilation	15
Temperature increase	>101.8°F	15
Cardiac involvement	Inappropriate sinus tachycardia	3

MH: Malignant hyperthermia

at few centers only, clinical grading scale for MH proposed by Larach can be useful in diagnosing a likely case of MH.

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#### Conflicts of interest

There are no conflicts of interest.

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