The Journal of International Medical Sciences Academy
2021; (April - June); Vol 34; No. 2

CASE REPORT

Malignant Hyperthermia during Scoliosis Correction.

Suresh S. Pillai[1], Sunil Paul Pathrose[2], Swaroop Sujath[3], Manikandan Maddy[3]

[1]Senior Consultant Spine Surgeon, [3]Orhtopedic Residents, Baby Memorial Hospital, Calicut, Kerala, India
[2]Consultant Spine Surgeon, Spine Centre, Keezhillam, Perumbavoor, Kerala, India

Abstract

A 14-year-old girl with severe rigid scoliosis and trunkal imbalance was planned surgical correction through a transthoracic anterior release and a second stage posterior instrumented correction and fusion. During the anterior release approximately 90 minutes after beginning of the surgery her body temperature rose from 36.8^{0}F to 105^{o}F and ETCO2 rose from 48 to 99 despite adjustments in the volumes. The variation was promptly picked up by the anesthetist. The vaporizer was removed, soda lime and ventilatory circuit was changed along with aggressive cooling measures in the form of per rectal and Riles tube cold saline instillation, covering the whole body with ice packs, and injection of cold saline intravenously. The patient settled soon with 100% oxygen application. She was ventilated for two more days after completion of the surgery. One week later her scoliosis was corrected through posterior instrumentation and fusion under total intravenous anesthesia (TIVA). She withstood the second surgery very well.

Keywords: **Malignant hyperthermia, Scoliosis correction, volatile anaesthetics, Total intravenous anaesthesis (TIVA), Dantrolene**

Introduction

Malignant hyperthermia is a life-threatening condition triggered by exposure to volatile anesthetic agents or neuromuscular blocking drugs like succinylcholine. A high index of suspicion and prompt monitoring with all modalities including temperature probe are imperative to safe a life. Dantrolene is a drug of choice for this condition, but it is not easily available. Here we report such a case of malignant hyperthermia in a patient who was undergoing scoliosis correction, with prompt recognition of the problem and immediate corrective measures resulting in an excellent outcome. The patient did not give any history of any immediate relatives being affected by this condition.

Case Report

A 14-year-old girl presented with truncal imbalance and shoulder imbalance secondary to severe scoliosis. She was diagnosed to have scoliosis at 8 years of age. She was evaluated with standing whole spine x-ray (antero-posterior and lateral views) and right and left supine side bending roentgenogram. She had Lenke Type 3 curve (Double Major) with Cobb angle measuring 100 degree from D3-L2 (right) and 60 degree from L2-L5 (left). The supine side bending films showed the thoracic curve correction from 100 to 80 degrees and the lumbar spine got corrected from 60 to 40 degrees. Her pulmonary function test showed restrictive and obstructive pattern. She weighed 25.5 kg with a BMI of 13.78. She had poor effort tolerance so the post-operative elective ventilation was explained. She was planned for right thoracotomy anterior release followed by second stage posterior instrumented correction and fusion of thoracic and lumbar curves a week after.

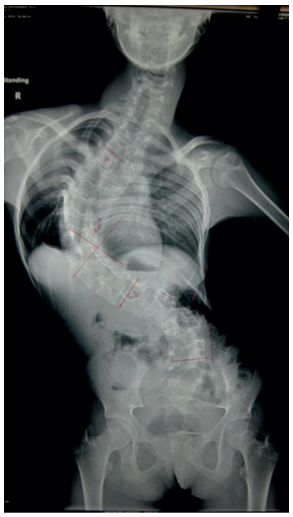

Figure 1: Preoperative standing radiograph

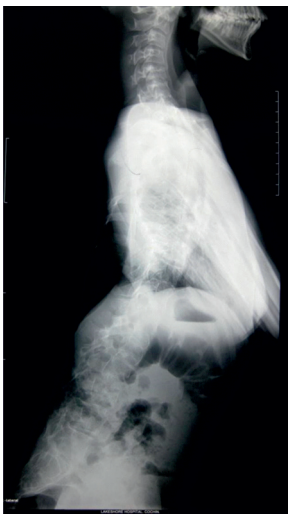

Figure 2: Preoperative standing lateral radiograph

Address for Correspondence
Dr. Suresh S. Pillai, Senior Consultant Spine Surgeon, Baby Memorial Hospital, Calicut, Kerala, India
E-Mail: sureshorth@gmail.com

Received: May 2020
Accepted: March 2021

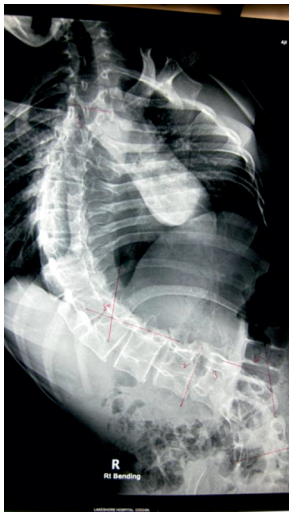

Figure 3: Preoperative supine side bending radiograph

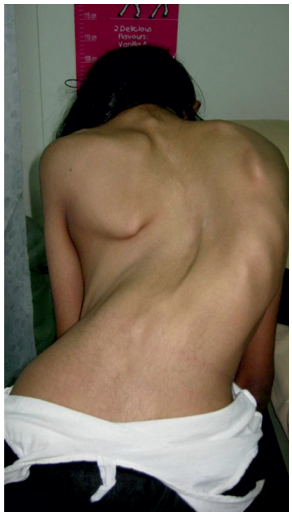

Figure 4: Preoperative clinical picture

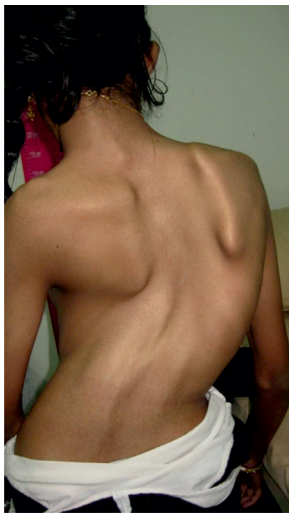

Figure 5: Pre-Operative standing clinical photograph

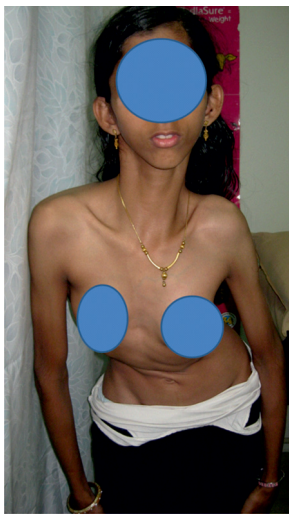

Figure 6: Preoperative standing-clinical photograph

The patient was pre-medicated by the anesthetist with Lorazepam 1 mg and Ondancetron 4 mg and induced by Glyco 1 mg, Fentanyl 50 mcg, Thyo 150 mg + Atracuriam 25 mg. Intubation done with a left-sided double lumen endotracheal tube (26 size). Right-sided internal jugular vein triple lumen CV catheter was placed and another arterial line on the left radial artery. Patient was monitored with ECG, IVP, SpO2, ETCO2, temperature, & urine output. She was maintained on oxygen plus nitrous oxide plus desflurane plus Atracuriam infusion and external body warming. Heart rate was 90 per minute and blood pressure 108/68 mmHg. Baseline ABG showed pH of 7.27, PCO2 49, PO2 139, & HCO3 22, & BE 5. Initial ETCO2 was 48, remained high, so OLV not done. A low tidal volume and increased frequency was given for the thoracotomy. A right thoracotomy was done sacrificing a rib at the apex. The collapsed lung on the right side was pushed away from the spine with mops. Six discs its annulus was removed around the apex, preserving the segmental vessels, to make the vertebral column supple. The resected rib was nibbled and used as the graft for anterior fusion. The parietal pleura was repaired. Halfway through the parietal pleural repair the anesthetist asked to stop the surgery as ETCO2 increased from 48 to 99 despite adjusting MV. She had tachycardia with heart rate increasing to 140 from 90 per minute. Her core temperature rose from 96.8F to 105.8F. ABG showed pH of 7.02, PCO2 98, PO2 131, HCO_3 24 and BE 10. The anesthetist asked to stop the surgery and turned the patient supine. The parietal pleura was not closed completely. With sterile towels over the wound the patient was turned supine. All the anesthetists in the hospital assembled there and discussed the plan of action quickly. They provisionally made a diagnosis of malignant hyperthermia. So, the patient was put on 100% oxygen, external warming stopped, and aggressive cooling started immediately. Ice cold saline was pushed through Ryle's per rectally. The entire body was covered with ice packs. Antipyretics were given. The vaporizer was removed. Soda lime and ventilatory circuit changed. Double lumen tube changed to single lumen endotracheal tube. Blood samples were sent for serum electrolytes and lactate. Soda bicarb 25 ml and calcium gluconate 10% 10 mL were given along with 50 mL of mannitol. Then ETCO2 reached 99 mmHg within 10 minutes of cooling and change of ventilatory circuit. Slowly, ETCO2 started coming down. The temperature also started declining from 105 to 100.4F in 20 minutes' time. The ABG at that point read pH of 7.22, PCO2 54, PO2 377, bicarb 23, and ABE 7. Patient was hemodynamically stable except for tachycardia. At this point the surgery restarted and completed. Then the patient was shifted to ICU for elective ventilation. Postoperatively, serum creatinine kinase was 1230 and on second day it was 1830 (less than 167 U/L), urine myoglobin was negative, serum myoglobin was 1073 ng/mL (25-58). Blood urea was 24 and serum creatinine was 0.6 mg%. Sodium 139, potassium 4.3, calcium 8.3, lactate 9 mg/dl postoperatively and 6 mg/dl on second postoperative day. Her INR was 1.43, D-dimer was negative. Serum creatinine kinase value normalized by fifth postoperative day. She was extubated the next day of surgery and was on intermittent NIV for two more days.

Posterior release and correction was planned two weeks after the first surgery. Anesthesia machine was prepared, dantrolene was not available, so TIVA was planned (fentanyl, Atracuriam, propofol, dexmeditomedine infusions, O2, nitrous oxide). Patient positioned prone, draped, and posterior release (bony & soft tissue) of thoracic and lumbar spine performed. Pedicle screws were used for correction. After correction, wake up test was performed. A paraspinal muscle biopsy was also performed. Local bone graft was used for fusion. She was electively ventilated on that day

and extubated the next day. Epidural infusion was given as postoperative analgesia. The patient had an uneventful recovery.

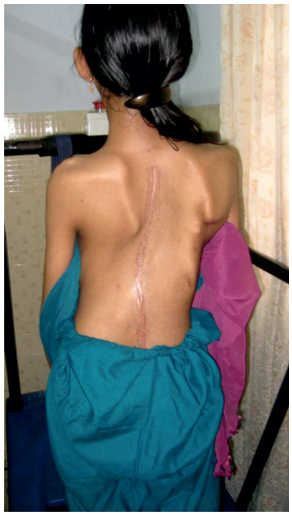

Figure 5: Pre-Operative standing clinical photograph

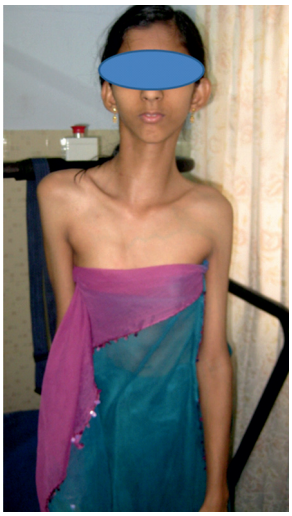

Figure 6: Preoperative standing-clinical photograph

Discussion

In the above setting the differential diagnoses considered were:

1. Anesthetic machine malfunction
2. Pheaochromocytoma
3. Thyroid crisis
4. Neuroleptic malignant syndrome

Malignant Hyperthermia

It is a rare life-threatening condition that is usually triggered by exposure to certain drugs used for general anesthesia, specifically the volatile anesthetic agents and the neuromuscular blocking agent, succinylcholine. MH has autosomal dominant genetic basis. 50% of the patients are caused by mutation on chromosome 19 in region that encodes the hydrophilic aminoterminal portion of the RYR1 receptor.

Malignant hyperthermia (MH) was first recognized in Australia in an affected family by Denborough et al in 1962. The efficacy of dandrolene was discovered by a South African anesthesiologist, Gaisford Harrison in 1975. The incidence of MH ranges from 1:5000 - 100,000 procedures involving general anesthesia. Majority of the affected people are children or young adults [1,2]. It is seen all across the world and affects all racial groups [3].

The pathophysiology of MH is thought to be a mutation encoding for abnormal RYR1 or DHP receptors that trigger unregulated passage of calcium from sarcoplasmic reticulum into the intracellular space. The accumulation of mycoplasmic calcium causes sustained muscle contraction, which over time generates heat. Accelerated levels of aerobic metabolism sustain the muscle temporarily, but produce carbon dioxide and cellular acidosis and deplete oxygen and ATP. A change to anaerobic metabolism worsens acidosis with production of lactate. Once energy levels are depleted the muscle fibers die, with rhabdomyolysis leading to hyperkalemia and myoglobinuria.

Six types of MH susceptibility (MHS 1-6) have been described based on the chromosomal locus affected.

MHS 1: Associated with RYR1 gene on chromosome 19q13.1

MHS 2: Associated with DHP receptor isolated to the 17q11.2 - q24 locus

MHS 3: Associated with the alpha 2/gamma sub unit of the DHP receptor, linked to 7q21-q22 locus

MHS 4: Linked to the 3q13.2 locus

MHS 5: Encoding of the alpha 1 sub unit of the DHP receptor and locus 1q32

MHS 6: Linked to chromosomal locus 5p

A few conditions are associated with MH. They are:

1. Myopathies (cenral core myopathy, multi-core myopathy, King Denborough syndrome, native American myopathy)
2. Rhabdomyolysis may be heat induced rhabdomyolysis, exercise-induced rhabdomyolysis, dystrophinopathies, myoadenylatedeaminase deficiency, McArdie's disease, carnitinepalmitoyltransferase type 2 deficiency)

RAW score is used to assess MH rank and qualitative likelihood

RAW Score Range	MH Rank	Description of Likelihood
0	1	Almost never
3-9	2	Unlikely
10-19	3	Somewhat less than likely
20-34	4	Somewhat greater than likely
35-49	5	Very likely
50+	6	Almost certain

MH susceptibility [4] can be tested using genetic test as well as caffeine-halothane contracture test. Contracture test is indicated for patients with a history of suspicious for MH, first-degree relatives of a patient with suspicious history. Patients with a suspicious history who are contemplating military service and others who have unexplained rhabdomyolysis following anesthesia, mild to moderate masseter muscle rigidity following succinylcholine administration, severe or recurrent exercise-or heat-induced rhabdomyolysis.

When giving anesthesia to MH susceptibility patients non-triggering agents are to be used. Prophylactic pharmacological intervention is not indicated. The anesthesia machine should be cleaned by flushing it with high-flow oxygen for 20 minutes.

Vaporizer canisters should be removed altogether. End-tidal carbon-dioxide (ETCO2) levels, minute ventilation, and core body temperature are monitored closely in these patients. Regional anesthesia is another option. All intravenous anesthetics and sedative agents like propofol, ketamine, etomidate, dexmeditomedine, barbiturates, or local anesthetics non depolarizing neuromuscular blockers, inhalational agents limited to nitrous oxide and xenon, analgesics, anxiolytics including opiods and benzodiazepines are safe agents in MH susceptible individuals.

Emhg Guideline (European MH Guidelines) 2010 [5]

Managing an MH crisis:

- Start treatment as soon as MH crisis is suspected.
- The clinical presentation of MH varies and treatment should be modified accordingly.
- Stop all trigger agents
- Hyperventilate (use minute volume 2-3 times normal) with 100% oxygen at high flow
- Call for help
- Change to non-trigger anesthesia (TIVA)
- Inform the surgeon and ask for termination/ postponement of surgery
- Disconnect the vaporizer - do not waste time changing the circuit/anesthetic machine
- Dantrolene in a dose of 2 mg/kg IV (ampules of 20 mg are mixed with 60 ml sterile water). Dantrolene infusion should be repeated until the cardiac and respiratory system stabilize. Maximum dose of 10 mg/kg.

EMHG on monitoring:

- Continue routine anesthetic monitoring (SAO2, ECG, NIBP, ETCO2)
- Measure core temperature
- Establish good IV line with wide bore cannula
- Consider inserting an arterial and central venous line and a urinary catheter
- Obtain samples for measurement of potassium, CK (creatinine kinase), arterial blood gases, myoglobin, and glucose
- Check renal and hepatic function and coagulation
- Check for signs of compartment syndrome
- Monitor the patient for a minimum of 24 hours (ICU, HDU, or in a recovery unit

Symptomatic Treatment

- Two to three liters of chilled (4-8° celcius) 0.9% saline IV
- Surface cooling: wet, cold sheets, fans, and ice packs placed in the axillae and groin
- Other cooling devices if available
- Stop cooling once temperature is 38.5° celcius
- Treat hyperkalemia with 50 ml of 50% dextrose with 10 IU insulin, calcium chloride 0.1 mmol/kg IV
- Dialysis may be required if the patient has myoglobinuria
- Acidosis may be treated by hyperventilation to normocapnoea and sodium bicarbonate IV if pH is less than 7.2
- Arrhythmias may be treated with amiodarone or beta blockers
- Maintain urinary output more than 2 ml/kg per hour
- Frusemide 0.5 to 1 mg/kg, manittol 1 gm/kg may be used
- Crystalloids (Ringer lactate solution or 0.9% saline IV)

Conclusion

A high index of suspicion for MH whenever you use volatile anesthetics and succinylcholine is required to prevent catastrophic events. Early detection and aggressive management can save life in MH [6,7]. Patients and family members of individual testing positive for susceptibility to MH should be counseled about this disorder

Conflict of interest:	All authors declare no COI
Ethics:	There is no ethical violation as it is based on voluntary anonymous interviews
Funding:	No external funding
Guarantor:	Dr. Suresh S. Pillai will act as guarantor of this article on behalf of all co-authors.

References

1. Can Jou Ana.1996-MH in a 10 yr old child manifested 10 min after starting desflurane.
2. AnesthAnalg.1997 -13 yr old boy for cerebral angiography after embolization of an arteriovenous malformation under GA with desflurane (90 min).
3. Uskova AA, Matusic BP, Brandom BW et al - Desflurane, malignant hyperthermia, and release of compartment syndrome - AnesthAnalg. 2005;100(5):1357-60
4. Glahn KP, Ellis FR, Halsall PJ, Müller CR, Snoeck MM, Urwyler A, Wappler F et al- Recognizing and managing a malignant hyperthermia crisis: guidelines from the European Malignant Hyperthermia Group - BJA 105(4):417-20(2010)-EMHG guidelines
5. Larach MG, Localio AR, Allen GC, Denborough MA, Ellis FR, Gronert GA, Kaplan RF, Muldoon SM, Nelson TE, Ording H, et al - A clinical grading scale to detect MH susceptibility - Anesthesiology 80:771-779,1994
6. IJA 2007-volume 51, issue:6-20year old for mandibular plating
7. The internet journal of pharmacology.2001, volume 1, 20 year old –following isoflurane