Malignant Hyperthermia Episode During Tethered Cord Release In A 4 Year Old Child

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Introduction: A 4 year old, ASA I, girl weighing 15.7 kg scheduled for tethered cord release had a past medical history significant for an abnormal gait, weak legs, and a previous uneventful general anesthetic with inhalational agents for bilateral myringotomies and tubes. There was no family history of any neuromuscular disease or adverse reactions to volatile anesthetic gases. Evaluation for neuromuscular disease was negative. MRI evaluation for scoliosis demonstrated a tethered cord.

Intraoperative Management: Anesthesia was induced with N₂O, O₂, Sevoflurane and tracheal intubation facilitated with rocuronium. Intravenous medications included glycopyrrolate, fentanyl, and cefazolin. The patient was positioned prone with equal and bilateral breath sounds. Volume controlled ventilation started with a peak inspiratory pressure (PIP) of 18 cmH₂O and an end-tidal carbon dioxide (EtCO₂) of 40 mmHg.

Surgical dissection revealed a tethered cord. Fifty-five minutes after induction, the EtCO₂ was noted to be gradually increasing. Light anesthesia, displaced endotracheal tube, mucous plugging, return of muscle tone, positioning, equipment malfunction, were quickly excluded and minute ventilation increased in response to increasing EtCO₂. Additional doses of fentanyl and rocuronium were administered, but the EtCO₂ continued to rise with a maximum value of 70 mmHg and a PIP of 22 cmH₂O. The patient became more tachycardic and developed muscle rigidity. Also, the esophageal temperature rose quickly to 38°C.

Malignant hyperthermia (MH) was suspected, the surgeon notified and MH protocol initiated. The anesthetic technique was converted to total intravenous anesthesia (TIVA) with propofol, fentanyl, and midazolam. Additional IVs and an arterial line were placed and dantrolene 40 mg IV administered resulting in the normalization of EtCO₂ and resolution of muscle rigidity. Table 1 The surgery was allowed to continue after some discussion. A urinary catheter was placed at the end of the procedure. The patient was awakened and the trachea extubated, after which she was transferred to the PICU in stable condition.

Postoperative Course: The PICU staff was informed of the MH episode and given the MH Hotline telephone number. It was determined the clinical grading score for MH was 58, making it almost certain this was a true event. The child continued to show evidence of a metabolic acidosis requiring dantrolene for another 48 hours. Urine output remained brisk for the duration of the PICU stay without evidence of myoglobinuria. The highest value for creatine kinase was 4824 u/L on the morning of surgery. The child had mild nausea and slight weakness in the postoperative period attributed to dantrolene. She was observed in the PICU for an additional 24 hours after the dantrolene was stopped and then transferred to the floor for further observation and counseling before being discharged on postoperative day 5.

Table 1

<table>
<thead>
<tr>
<th></th>
<th>pH</th>
<th>PCO₂</th>
<th>PaO₂</th>
<th>HCO₃</th>
<th>Base excess</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pre Dantrolene</td>
<td>7.21</td>
<td>54</td>
<td>119</td>
<td>21.2</td>
<td>-6.8</td>
</tr>
<tr>
<td>Post Dantrolene</td>
<td>7.25</td>
<td>53</td>
<td>212</td>
<td>23</td>
<td>-4.2</td>
</tr>
</tbody>
</table>

Discussion: Our case report presents several unique dilemmas. The onset of malignant hyperthermia happened late into the case. In a previous study, the late onset of MH has correlated with a more prolonged recovery period. Our child manifested clear signs of MH approximately one hour after exposure to anesthetic gases and required dantrolene for an additional 48 hours. The early intervention appears to have aborted the MH episode quickly with minimal tissue damage, a peak creatine kinase level of 4824 u/L with no detected myoglobinuria. The response to dantrolene was dramatic and as the procedure was near to completion we decided to continue the operation. Body temperatures above 40°C predict the occurrence of multi-organ system failure with DIC, which is uniformly fatal. Our patient was alert, hemodynamically stable, had a brisk urine output and did not manifest any signs or laboratory evidence of DIC. Early recognition and prompt therapy with dantrolene may be responsible for the rapid response in our case. Finally, the decision to electively extubate the child was based on the rapid clinical response to dantrolene, at the same time being aware of the potential side effects of dantrolene.

References:

3) Burkman, JM, Posner KL, PhD, Domino KB  Analysis of the clinical variables associated with recrudescence after malignant hyperthermia reactions. Abstract Presentation, MHAUS, October, 2005