SUXAMETHONIUM ASSOCIATED HYPERTONICITY AND CARDIAC ARREST IN UNSUSPECTED PSEUODOHYPERTROPHIC MUSCULAR DYSTROPHY

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SUMMARY

We report a case that showed many of the features of the malignant hyperpyrexia syndrome following suxamethonium administration, in an unsuspected case of pseudohypertrophic (Duchenne) muscular dystrophy.

In 1971 a patient with suxamethonium-induced cardiac arrest in an unsuspected case of pseudohypertrophic muscular dystrophy was described (Genever, 1971). Suxamethonium has been implicated in several cases of muscular rigidity, hyperkalaemia and cardiac arrest and in malignant hyperpyrexia, and there is evidence that these reactions occur often when there are pre-existing muscular disorders including muscular dystrophies, hernias (Ellis and Halsall, 1980) and strabismus (Tammisetoa et al., 1970). The mechanisms are unknown.

A second patient with cardiac arrest in unsuspected pseudohypertrophic (Duchenne) muscular dystrophy is reported in this paper. In this case, however, there was a significant difference in that cardiac arrest was preceded by rigidity, cyanosis and acidosis.

CASE HISTORY

A male, only child, who had not walked until the age of 2 yr, but who had been healthy and considered normal by his parents, was admitted to hospital at the age of 5 yr for surgical correction of his right convergent squint which he had developed at the age of 2 yr. He had not been investigated for his delay in walking.

The patient was premedicated with papaveretum 5 mg and hyoscine 100 µg. After induction of anaesthesia with nitrous oxide, oxygen, cyclopropane and halothane he was given suxamethonium 25 mg i.v. and immediately developed generalized hypertonicity, such that it was difficult to open his mouth and, after intubation of the trachea, to inflate the lungs. Despite ventilation with 100% oxygen the patient became progressively more cyanosed with bradycardia, and seven minutes after the suxamethonium there was cardiac arrest. Resuscitation from asystole included treatment with i.v. sodium bicarbonate and adrenaline followed by intracardiac adrenaline and calcium gluconate before defibrillation for ventricular fibrillation resulted in sinus rhythm.

Thirty minutes after the intracardiac injection and about forty minutes after the suxamethonium the patient was rousable and the heart was in sinus rhythm with multifocal ventricular ectopics. The serum potassium concentration was 5.4 mmol litre⁻¹ and oral temperature 36.5°C. At no time did the patient appear hot, nor did his temperature increase.

Serial potassium estimations over the next seven days revealed values always less than 4.4 mmol litre⁻¹, and subsequent e.c.g. showed a normal trace with a tachycardia for the first three days. Serial arterial blood analysis showed a persistent acidosis.

Six hours after the cardiac arrest the patient was alert, orientated and talking. On examination of the lungs there were audible ronchi on the left side. Over the next twelve hours the patient’s respiratory effort decreased and he became unable to cough and clear his secretions, developing supraventricular tachycardia of 220 beat min⁻¹ which was resistant to digoxin. The lungs were ventilated artificially for four days, during which time the patient was given sodium bicarbonate and potassium by infusion to correct the acidosis as indicated (see table I).

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TABLE I. Administration of sodium bicarbonate to correct acidosis

<table>
<thead>
<tr>
<th>Time after arrest (h)</th>
<th>pH (unit)</th>
<th>Bicarbonate administered (mmol)</th>
<th>Heart rate (beat min⁻¹)</th>
</tr>
</thead>
<tbody>
<tr>
<td>0.5</td>
<td>7.17</td>
<td>12.5</td>
<td>220</td>
</tr>
<tr>
<td>20</td>
<td>7.21</td>
<td>20</td>
<td>160</td>
</tr>
<tr>
<td>28</td>
<td>7.20</td>
<td>20</td>
<td>150</td>
</tr>
<tr>
<td>31</td>
<td>7.27</td>
<td>20</td>
<td>160</td>
</tr>
<tr>
<td>38</td>
<td>7.28</td>
<td>23</td>
<td>150</td>
</tr>
<tr>
<td>48</td>
<td>7.41</td>
<td>175 ml x 20 per min</td>
<td>150</td>
</tr>
<tr>
<td>72</td>
<td>7.40</td>
<td>115</td>
<td>130</td>
</tr>
<tr>
<td>96</td>
<td>7.36</td>
<td>90</td>
<td>90</td>
</tr>
<tr>
<td>120</td>
<td>7.46</td>
<td>105</td>
<td>90</td>
</tr>
<tr>
<td>142</td>
<td>7.43</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Frequent chest physiotherapy was applied and physiotherapy was also required for progressive foot drop. The calves became progressively hypertrophic.

Serum creatinine phosphokinase on the second day was 30 000 i.u. and on day ten was 10 000 i.u. After recovery the patient had a typical Duchenne walk and was able to rise from sitting on the floor only by climbing up his legs. The diagnosis was confirmed by muscle biopsy and e.m.g.

**DISCUSSION**

Suxamethonium has previously been reported as being associated with cardiac arrest in pseudohypertrophic muscular dystrophy and prolonged hypertonicity in other cases.

The interesting feature of this patient was the immediate hypertonicity following injection of suxamethonium with a rapidly progressing cyanosis leading to cardiac arrest. This, together with the developing acidosis, brought to mind a possible diagnosis of malignant hyperpyrexia, although the temperature was never greater than 36.5 °C.

The mechanism of the cardiac arrest will remain in doubt. There is a possibility that the arrest was a result of hyperkalaemia, although the highest recorded serum potassium value was 5.4 mmol litre⁻¹, or the arrest may have been precipitated by hypoxia following some seven minutes after the rigidity and ensuing cyanosis. Duchenne muscular dystrophy is itself associated with heart defects, but in this case both e.g. and cardiac shadows on x-ray were normal. There was, however, a persistent supraventricular tachycardia for 72 h.

The muscle cell abnormality both in muscular dystrophy and malignant hyperpyrexia is unknown and this case shows many of the features of the malignant hyperpyrexia syndrome. Perhaps this should serve as a further warning that muscle relaxants may be hazardous in patients with any form of muscle disorder, including squint and hernia. A history of delayed motor milestones should also be treated seriously, as an indicator of underlying disorder, and thoroughly investigated.

**REFERENCES**


**NACH SUXAMETHONIUM AUFTRETENDER HYPERTONUS UND HERZSTILLSTAND BEI NICHT BEKANNTER PSEUDOHYPERTROPHISCHER MUSKELDYSTROPHIE**

Wir berichten über einen Fall, der viele merkmale der Malignen Hyperthermie aufwies, und nach Verabreichung von Suxamethonium bei einem Fall von nicht bekannter pseudohypertrophischer Muskeldystrophie (Duchenne).

**SUUXAMETONIO ASOCIADO CON HIPERTONICIDAD Y PARADA DEL CORAZÓN EN DISTROFÍA MUSCULAR SEUDO-HIPERTROFICA INSOSPECHADA**

Informamos sobre un caso en que se manifestaron muchos de los rasgos del síndrome de hiperpirexia maligna a raíz de la administración de suxamethonio en un caso insospechado de distrofia muscular seudo-hipertrofica (Duchenne).