Pseudocholinesterase Deficiency

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Recommended Citation
Gerken, Ross, "Pseudocholinesterase Deficiency" (2014). Master of Science in Nursing (MSN) Student Scholarship. 56.
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PSEUDOCHOLINESTERASE DEFICIENCY
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Introduction
Pseudocholinesterase deficiency is a rare autosomally inherited disorder typified by the metabolism of cholinesterase such as the neuromuscular blockers succinylcholine, pancuronium, and other local anesthetics. Pseudocholinesterase deficiency is an inherited genetic transmission, and an autosomal recessive pattern of inheritance. The pseudocholinesterase between 1:480 and 1:2000 people (Ok et al., 2013). An extended period of neuromuscular blockade results from these medications. What is currently expected is the signs and symptoms which occur during anesthesia, and not infrequently longer. This condition is rare but must be known and understood in the clinician to provide the safest patient care possible. The information is not widely known or readily available and usually is undetected until the patient receives the medication and then the neuromuscular blockade continues well beyond the expected duration. The purpose of this discussion is to further educate the practitioner on the pathophysiology of pseudocholinesterase deficiency, as well as signs and symptoms, and patient management strategies to improve patient outcomes.

Case Study
A 43 y/o female is having a routine c-section for a scheduled pregnancy. She has no known drug allergies. Her past medical history includes asthmatic bronchitis, which she takes Fluticasone propionate and 2.5mg montelukast. Past surgical history includes a left ankle fracture and a rotator cuff repair s/p rotator cuff tear. Her surgical history is one C section. She takes 20mg of trazodone at bedtime for a history of insomnia. She is allergic to penicillin. Her medications are usually hydrolyzed very quickly, even before it reaches the neuromuscular junction. According to Ok et al. (2013), “this hydrolysis rapidly inactivates around 90% to 95% of the intravenous succinylcholine dose. By contrast, to achieve complete hydrolysis and subsequent release of paralysis, the muscle paralysis must be maintained for up to 8 hours. After further examination vital signs were stable, blood glucose was 108, and electrolytes were within normal limits. However, train of four was assessed resulting in 4/4 twitch, indicating prolonged paralysis. This led to the care conclusion of Pseudocholinesterase deficiency. The major signs and symptoms occurred postoperatively after administration of the muscle relaxant succinylcholine for duration of surgery. The patient presented with muscle weakness, prolonged paralysis, and weak or 0/4 twitches with the train of four.

Pathophysiology
Pseudocholinesterase is an enzyme produced in the liver. The heart, plasma, pancreas and specific tissues matter also contain pseudocholinesterase Ama et al. (2010). The enzyme is responsible for metabolizing succinylcholine, mivacurium, and other othet local anesthetics such as succinates and procaines. In people with normal pseudocholinesterase levels, the medications are usually hydrolyzed very quickly, even before it reaches the neuromuscular junction. According to Ama et al. (2010), “this hydrolysis rapidly inactivates around 90% to 95% of the intravenous succinylcholine dose.

Dibucaine Inhibition Test Results

<table>
<thead>
<tr>
<th>Test</th>
<th>Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>Low dibucaine number</td>
<td>90% +/– 10% lower activity</td>
</tr>
<tr>
<td>Normal dibucaine number</td>
<td>Low activity with low levels present &amp; high levels present</td>
</tr>
<tr>
<td>Low number</td>
<td>Very low activity with very low levels present</td>
</tr>
<tr>
<td>Normal number</td>
<td>Same activity with same levels present</td>
</tr>
</tbody>
</table>

Dibucaine is an amide local anesthetic and is a competitive non-depolarizing muscle relaxant. Dibucaine is added to a sample of the patient’s serum and compared to untreated serum to show the sensitivity to succinylcholine. A Dibucaine number and enzyme activity are both determined. A normal result is 80. The results indicate Dibucaine inhibits 80% of pseudocholinesterase. A Dibucaine number of 20 is diagnostic for atypical pseudocholinesterase levels.

Disease Test
Disease test is a sample of the patient’s serum and compared to untreated serum to show the sensitivity to succinylcholine. A Dibucaine number and enzyme activity are both determined. A normal result is 80. The results indicate Dibucaine inhibits 80% of pseudocholinesterase. A Dibucaine number of 20 is diagnostic for atypical pseudocholinesterase levels.

Drugs Effects
- Baseline anesthetic, including over sedation and large amounts of narcotics.

Pathophysiology Cont.

<table>
<thead>
<tr>
<th>Cause</th>
<th>Effect</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypoglycemia</td>
<td>Hyperglycemia</td>
</tr>
<tr>
<td>Ischemic episode</td>
<td>Hypoxia</td>
</tr>
</tbody>
</table>

Conclusion
Pseudocholinesterase deficiency is a rare cause of prolonged paralysis. The potentiality for this deficiency can be life threatening, thus indicating the importance of education about this topic. A thorough prospective evaluation is crucial including a detailed family history and always further assessing a patient who states a family history or personal history of difficulty waking up after surgery. The clinician must be prepared to stay alert and responsive to the patient's presentation of symptoms, which is prolonged apnea after surgery with a neuromuscular blocking agent.

References


