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Pseudocholinesterase Deficiency: Implications in Anesthesia

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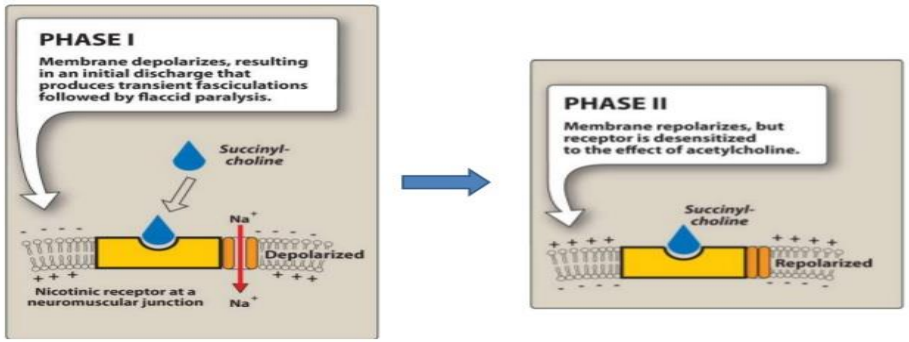
Introduction

The use of neuromuscular blocking medications is a frequent and essential practice in establishing a secure airway in patients preparing for surgery or in emergent situations such as respiratory distress. As an anesthesia provider, it is vital to know the mechanism of action, expected duration, and metabolism of these neuromuscular blocking agents particularly succinylcholine. A potential serious complication that may occur in patients that have been administered succinylcholine is prolonged neuromuscular blockade due to the deficiency of pseudocholinesterase, the enzyme produced in the liver and present in plasma that metabolizes succinylcholine (Branch, Rafacz, & Boudreaux, 2011). In patients with a pseudocholinesterase deficiency, succinylcholine is not broken down as quickly as would be expected, which produces a prolonged neuromuscular block. These patients will need continuous ventilator support post-operatively and often admission to the intensive care unit until muscle strength is restored (Stoelting & Hiller, 2015). Identifying those patients at risk and recognizing the signs of pseudocholinesterase deficiency are key in avoiding complications associated with an abnormally prolonged neuromuscular block.

Pathophysiology

- Pseudocholinesterase is an enzyme produced in the liver that metabolizes neuromuscular blocking agents, particularly succinylcholine and mivarcurium, as well as some local anesthetics.
- Pseudocholinesterase hydrolyzes succinylcholine in the plasma so quickly that only 10% of the medications actually reaches and acts upon the neuromuscular junction (NMJ).
- Once at the NMJ, succinylcholine binds to acetylcholine receptor sites causing sustained depolarization for a relatively short period of time in which the patient experiences skeletal muscle fasiculations followed by flaccid paralysis.
- Succinylcholine diffuses away from the NMJ down its concentration gradient at which point muscle function will return.
- Typically, an intubating dose of succinylcholine is rapidly hydrolyzed by pseudocholinesterase within 9 to 13 minutes in which 90% of the muscle strength will be restored, however its duration of action will be prolonged in a deficient patient depending on the type of genetic abnormality present.
- Pseudocholinesterase deficiency is caused by either a homozygous or heterozygous abnormal genetic variant of the enzyme itself. An individual that is homozygous for the atypical gene which occurs in one in every 3,500 people, the neuromuscular block produced by succinylcholine will likely last four to eight hours and this genotype is more likely to experience apnea and the inability to be taken off of the ventilator post-operatively.
- An individual with a heterozygous gene which occurs one in every 480 people, will experience the effects of succinylcholine two times longer than an individual with a normal genotype.
- There are multiple other causes that are less significant, but may cause a decrease in pseudocholinesterase activity including liver disease, old age, pregnancy, burns, malnutrition, neoplastic disease, the use specific mediations such as reglan, oral contraceptives, monamine oxidase inhibitors, and anticholinesterase medications (Stoelting & Hiller, 2015).



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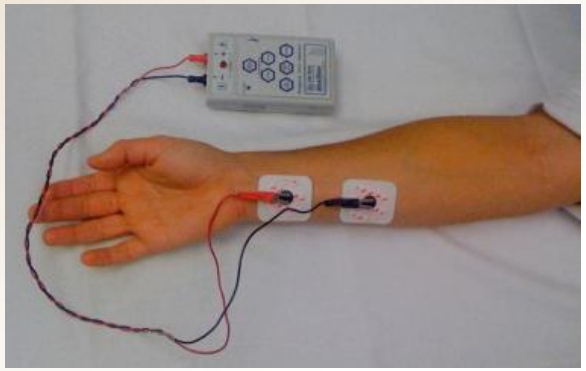
Significance of Pathophysiology

- While pseudocholinesterase deficiency is not one of the most common complications seen with general anesthesia, it can be fatal if not recognized and treated properly.
- Since we know there is a genetic link, a thorough pre-operative assessment should be completed including family history of this condition. Sometimes asking if the patient or a family member was admitted to the ICU or had to be on the ventilator for longer than expected after surgery will help to reveal a pseudocholinesterase deficiency.
- The deficiency is most often found after succinylcholine has already been given and it is discovered that the patient's muscle function has not returned within the expected time frame.
- There is no treatment for this genetic condition, however supportive care, particularly ventilator support, must be given until the medication has been adequately metabolized and the patient has regained muscle strength (Stoelting & Hiller, 2015).

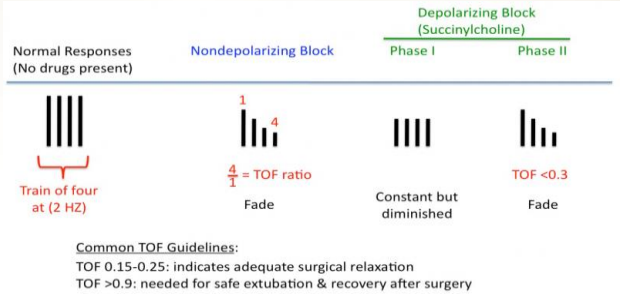


Signs and Symptoms

- A patient with pseudocholinesterase deficiency will not experience signs or symptoms unless he or she is administered a neuromuscular blocking agent.
- The major presenting sign is prolonged skeletal muscle paralysis including the diaphragm and intercostal muscles that are required for the patient to breath.
- Apnea and a lack of movement or muscle twitches when using a peripheral nerve stimulator will present until the medication is metabolized by the body which may not be until hours later (Whittington, et. al, 2012).
- The peripheral nerve stimulator is a method for monitoring neuromuscular blockade in which electrodes are placed either on the facial or ulnar nerve and stimulated using various modes to produce a muscle twitch. Train-of-four is a commonly used mode in which four simultaneous stimuli are delivered and the number and strength of the tissues is observed by the anesthesia provider. The goal after using a neuromuscular blocking agent and prior to extubation is four twitches without fade which refers to the strength of the twitch. An individual with a pseudocholinesterase deficiency may not produce any twitches depending the amount of time that has passed since administration and the type of genetic variant (Nagelhout, & Plaus, 2013).

Implications for Nursing Care

- Nursing anesthesia providers to know the mechanism of action and potential adverse effects of all medications especially neuromuscular blockers in order properly recognize and treat a pseudocholinesterase deficiency.
- Always assess muscle function using train of four monitoring with a peripheral nerve stimulator prior to an after a neuromuscular blockade is given. This will allow for comparison and the provider can be more confident in the degree of muscle strength recovery prior to making the decision to extubate the patient (International Anesthesia Research Society, 2016).
- Do not extubate a patient that has not experienced muscle recovery, provide full ventilator support, and make arrangements for a ventilator set-up in the PACU or ICU.
- Avoid the use of succinylcholine unless absolutely necessary and do not give unless the benefits outweigh the potential costs (Quyen, 2012).
- PACU and ICU nurses should familiarize themselves with the significance of a pseucholinesterase deficiency, the use of a peripheral nerve stimulator, and the necessary care for these patients postoperatively.



Retrieved from http://tmedweb.tulane.edu/pharmwiki/doku.php/nicotinic_antagonists

Conclusion

- This condition will not affect the patient's everyday life, however it may become a life-threatening condition if the patient must undergo general anesthesia for surgery.
- For this reason anesthesia providers should always consider using less invasive airway options or nerve blocks, both of which would not require the administration of neuromuscular blocking agents, like succinylcholine.
- If the use of a NMB cannot be avoided and there are no absolute contradindications, then providers must be prepared to recognize, assess and provide support to a patient that presents with a pseucholinesterase deficiency.

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Additional Resources

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