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Pseudocholinesterase Deficiency

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PSEUDOCHOLINESTERASE DEFICIENCY

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Introduction

Pseudocholinesterase deficiency is a rare genetic or acquired variation in the metabolism of choline esters such as the neuromuscular blockers succinylcholine, mivacurium, and ester local anesthetics. Pseudocholinesterase deficiency genetically is transmitted in an autosomal recessive pattern with the frequency of apnea from a genetic abnormality of pseudocholinesterase between 1:480 and 1:3200 people (Ok et al., 2013). An extended period of neuromuscular blockade results from these medications than what is clinically expected. The signs and symptoms which occur are apnea and paralysis hours longer. This condition is rare but must be known and understood by the clinician in order 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 medications 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 year old female is having rotator cuff repair s/p rotator cuff tear. She has no known drug allergies. Her past medical history is GERD and hypertension, which she takes 20mg lisinopril and 20mg omeprazole. Past surgical history is one C-section. Family history of anesthetic problems: States "thinks her dad may have had difficulty waking up but was not sure". Vitals: blood pressure 128/86, heart rate 70, SpO2 99% on room air, respirations 16, temperature 98.4°C. The patient had general anesthesia, with standard induction consisting of 80mg lidocaine, 150mg propofol, 5mg rocuronium, and 140mg succinylcholine. Sevoflurane 2% was used for maintenance. Intra-operative course is uneventful. During emergence patient remains apneic, with no response to increase in CO₂. First considerations must be differential diagnoses for a delayed emergence.

Differential Diagnosis

Drug Effects:

- Residual anesthetic, including over sedation and large amounts of narcotics.

Metabolic causes:

- Hypothermia
- Hypoglycemia
- Hyponatremia
- Hypoxia
- Hypercarbia

Neurological causes:

- Increased cerebral hemorrhage resulting in increased intracranial pressure
- Ischemic episode
- Seizure resulting in a post-ictal state

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 0/4 twitches, indicating prolonged paralysis. This lead to the rare conclusion of Pseudocholinesterase deficiency. The major signs and symptoms occurred postoperatively after administration of the muscle relaxant succinylcholine for induction during surgery. The patient presented with apnea, 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 the white matter also contain pseudocholinesterase Ama et al. (2010). The enzyme is responsible for metabolizing succinylcholine, mivacurium, and other ester local anesthetics such as cocaine and procaine. 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.

Pathophysiology Cont.

The remaining 5% to 10% of the succinylcholine dose acts as an acetylcholine receptor agonist at the neuromuscular junction." This causes depolarization of skeletal muscle, which results in fasciculations that can be seen in the muscles. Once the acetylcholine is released from the presynaptic membrane, and the remainder of the succinylcholine binds to the receptors on the postsynaptic neuromuscular junction, it is no longer able to evoke any more action potentials. This is what causes the muscle paralysis. The paralysis can develop as quickly as one minute. Elimination of succinylcholine is accomplished by pseudocholinesterase enzyme diffusing the medication away from the acetylcholine receptors. It is first metabolized into succinylmonocholine, which is 1/20th to 1/90th as potent as succinylcholine and then broken down to succinate and choline Ama et al. (2010). Normally, a patient recovers fully from succinylcholine (0.5-1.0 mg/kg) within 5 minutes. However, a patient who has pseudocholinesterase deficiency is unable to metabolize the medication as quickly as a person with normal levels. This leads to increased number of succinylcholine molecules in the neuromuscular junction, causing the duration of paralytic effect to continue for up to 8 hours.

Dibucaine Inhibition Test Results

Nursing Implications

Nursing management of prolonged paralysis due to plasma cholinesterase deficiency	
Nursing skill	Rationale
Know effects of neuromuscular blocking agents	Depolarizing neuromuscular blocking agents (eg, succinylcholine) <ul style="list-style-type: none">• Drug directly attaches to the postsynaptic acetylcholine receptor at the motor end plate, causing persistent depolarization of motor end plate. Drug effects wear off as plasma cholinesterase enzyme rapidly hydrolyzes the drug. No reversal agent exists. Nondepolarizing neuromuscular blocking agents (eg, mivacurium) <ul style="list-style-type: none">• Drug blocks the acetylcholine receptor and prevents binding to the postsynaptic acetylcholine receptor. Drug prevents depolarization of plasma membrane of motor end plate and muscle contraction. Effects wear off after the drug is hydrolyzed.
Use peripheral nerve stimulator	Quantify train of 4 to determine degree of blockade: 4 twitches=0%-75% blockade 3 twitches=80% blockade 2 twitches=85% blockade 1 twitch=90% blockade 0 twitch= 100% blockade
Administer sedative/amnestic	Reduce the patient's anxiety, stress, fear, or loss of physical function during paralysis
Assess resolution of neuromuscular blockade, sedation, and readiness for extubation	Anticipate risk of residual neuromuscular blockade, poor muscle strength, and premature extubation. Assess for the following: score of at least -1 or 0 on Richmond Agitation Sedation Scale, return of gag/cough reflex, ability to lift head off the pillow for ≥5 seconds, ability to lift both legs off the bed for ≥5 seconds, ability to maintain bilateral hand grips for ≥5 seconds, consistent and adequate tidal volumes and respiratory rate on ventilator pressure settings.
Recognize abnormal results of laboratory tests	Obtain venous blood sample 24 hours after administration of neuromuscular blocking agent to quantify plasma cholinesterase level and dibucaine inhibition number. Low plasma cholinesterase level means decreased ability to hydrolyze succinylcholine or mivacurium. A decreased dibucaine inhibition number (<80%) indicates the presence of an abnormal plasma level of cholinesterase. A genetic consultation should be considered.
Take steps to prevent complications and to ensure patients' safety	Conduct thorough preoperative assessments, flag medical records that indicate adverse reaction to any medication, especially anesthetics. Provide interventions to prevent skin breakdown, decrease risk for ventilator-associated pneumonia, and avoid impaired functional mobility after extubation. Implement teaching plan for patient to be knowledgeable about implications of plasma cholinesterase deficiency.

Dibucaine Inhibition Test Outcomes

1. Low dibucaine number + slightly lower activity = atypical enzyme & prolonged apnea
2. Normal dibucaine number + low activity = normal enzyme with low levels present & prolonged apnea
3. Low dibucaine number + very low activity = possible rare variant-type enzyme with very low levels present and prolonged apnea
4. Normal dibucaine number + normal activity = normal enzyme and amount (other causes of prolonged apnea must be investigated)

Diagnostic Test:

Dibucaine is an amide local anesthetic that inhibits typical pseudocholinesterase. 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 activity. A Dibucaine number of 20 is diagnostic for atypical pseudocholinesterase levels.

Treatment

Treatment is aimed at patient safety, care and comfort. The standard of care is to maintain sedation and ventilation until return of normal neuromuscular function and patient is able to protect their airway. This is considered the safest option. The other options are transfusions of whole blood and FFP, or purified human cholinesterase.

Conclusion

Pseudocholinesterase deficiency is a rare cause of prolonged paralysis. The potential risk of this deficiency can be life threatening, thus indicating the importance of education about this topic. A thorough preoperative 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 aware of the most recognizable symptom, which is prolonged apnea after surgery with administration of the neuromuscular blocker succinylcholine.

Conclusion Continued

The most important issue is to keep the patient safe and comfortable throughout this experience. Always remember just because the patient is paralyzed does not mean they are not aware or pain free. The patient must be kept sedated with adequate pain control until the prolonged paralysis resolves.



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