Pseu do cho lin es ter ase deficiency is an inherited or acquired condition in which the metabolism of suc cinylcholine, mivacurium, or ester local anesthetics is potentially impaired. In this review, genetic inheritance, variants, and testing are examined. Additionally, acquired conditions and drugs that influence enzyme activity, as well as possible treatments of the condition, are reviewed.

The review of the literature was conducted by searching PubMed and Ovid Medline databases, with no limitation on date of publication. The search was limited to English-language journals only. Additional articles of relevance were obtained from reference lists of previously searched articles and via Internet searches. Numerous keywords were used in the search, and a second search was undertaken to find specific citations about acquired conditions and drugs of relevance. Nearly 250 articles were obtained and examined for importance. Fifty articles appear in the review, including case reports, research studies, and review articles.

**Keywords:** Atypical pseudocholinesterase, BChE, butyrylcholinesterase, genetic variants, pseudocholinesterase.

**Butyrylcholinesterase**—also known as pseudocholinesterase, serum cholinesterase, plasma cholinesterase, and false cholinesterase—was so named due to its ability to hydrolyze butyrylcholine faster than other esters. Pseudocholinesterase is produced in the liver and found in most tissues, with the exception of red blood cells. The presence of pseudocholinesterase in the body has been established for well over a century. Although potential functions of this enzyme are debated to this day, its role in the metabolism of choline esters is universally recognized.

**Definition of Pseudocholinesterase Deficiency**
Pseudocholinesterase deficiency is a genetic or acquired alteration in the metabolism of choline esters such as succinylcholine, mivacurium, and ester-linked local anesthetics. The most described consequence of pseudocholinesterase deficiency in the literature is prolonged paralysis and apnea after administration of succinylcholine or mivacurium. The latter drug is no longer produced in the United States but is used elsewhere in the world. Less frequently described are adverse outcomes with the use of ester local anesthetics, particularly chloroprocaine.

Individuals can live their entire lives with pseudocholinesterase deficiency and not experience any untoward health effects, and the presence of the genetic defect is not realized until one is exposed to succinylcholine or mivacurium. In fact, a pseudocholinesterase-deficient group of individuals in the Vysya community of India were studied, and the determination was made that when individuals were not challenged with drugs or poisons, and did not consume a high-fat diet, the absence of butyrylcholinesterase enzyme caused no adverse health effects.

**Relevance to Anesthesia Providers**
Pseudocholinesterase deficiency, whether inherited or acquired, is an important concept to understand for providers that administer succinylcholine, including anesthesia, intensive care unit (ICU), emergency department, and perioperative personnel. We must appreciate basic concepts of genetics, genetic testing, and pharmacogenetics to make informed choices about our plan of care. We must also be able to extract what diseases, conditions, or medications would cause alterations in a patient’s enzyme activity and make thoughtful choices about our care based on this information.

The aim of this review is to examine both the genetic aspects of pseudocholinesterase deficiency and the acquired aspects to help the reader stay abreast of established and current literature and theories.

**Genetics of Pseudocholinesterase Deficiency**
Refer to Table 1 for a list of common genetic terms used throughout this article.

- Inheritance of Atypical Pseudocholinesterase or Pseudocholinesterase Deficiency. Genetically inherited pseudocholinesterase deficiency is typically regarded as an auto-
somal recessive trait, although it does not adhere to the traditional definition completely. Pseudocholinesterase deficiency is associated with the butyrylcholinesterase or BCHE gene and is located on the long arm of chromosome 3 at 3q26.1-26.2. It is a relatively short piece of genetic material, making DNA testing for the deficiency fairly feasible.

- **Variants.** There are 65 named variants one can inherit that may result in minimal to extreme postsuccinylcholine apnea and paralysis.\(^3\) The most frequently discussed and possibly the most clinically relevant are the atypical or dibucaine resistant, the fluoride resistant, the silent, and the K variants. Refer to Table 2 for a list of discussed variants, frequencies, and anesthetic implications after succinylycholine administration.

- **Atypical or Dibucaine Resistant.** The existence of a hereditary link for pseudocholinesterase deficiency had been speculated on in the literature for decades. In the mid-20th century the pioneering work by Kalow and colleagues\(^4,5\) showed the presence of a qualitative difference in plasma cholinesterase enzymes of sensitive families compared with the remainder of the population. They later introduced the dibucaine inhibition test.

Thus, the first discovered and discussed variant was the atypical or dibucaine resistant. The local anesthetic dibucaine was used as an inhibitor, and benzocholine was used as a substrate to determine the activity of pseudocholinesterase. The term dibucaine number was coined, meaning the percent inhibition of enzyme activity by dibucaine when a serum or plasma sample is tested under standard conditions. A dibucaine number above 70 is described as typical or normal, a number between 40 and 70 as intermediate (heterozygotes), and a dibucaine number below 20 as atypical (homozygotes).\(^4,5\) An individual with a normal dibucaine number would react normally to succinylcholine, a heterozygote would generally be expected to have a normal or minimally prolonged response to succinylcholine, and homozygotes would be expected to have postsuccinylcholine apnea and paralysis lasting near 2 hours or longer. Frequencies of inheritance vary by population, but generally heterozygotes can account for 1 in 25 to 1 in 50 individuals, whereas homozygotes can account for 1 in 3,000 individuals.\(^6\)

- **Fluoride Resistant.** Shortly after the classification of variants using the dibucaine number, Harris and

### Table 1. Definitions of Common Genetic Terms

<table>
<thead>
<tr>
<th>Term</th>
<th>Definition</th>
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<tbody>
<tr>
<td>Chromosome</td>
<td>Unit of DNA that contains genes. There are normally 23 paired chromosomes (46 total chromosomes per cell). Consists of 2 arms: the p arm, which is the short arm, and the q arm, the long arm.</td>
</tr>
<tr>
<td>Gene</td>
<td>Occupies a specific area on a chromosome. Consists of DNA and contributes both to the characteristics of an organism and specific protein coding.</td>
</tr>
<tr>
<td>Wild type/typical/normal</td>
<td>Most common version of the gene</td>
</tr>
<tr>
<td>Variant</td>
<td>Different version of the gene, with or without consequences to the individual</td>
</tr>
<tr>
<td>Allele</td>
<td>Alternate form of a gene occupying a specific area on a chromosome</td>
</tr>
<tr>
<td>Homozygous</td>
<td>Two copies of same allele. Using pseudocholinesterase deficiency as an example, a normal individual would be considered UU—inherting the same wild-type allele from both parents—and an individual homozygous for the atypical variant would be considered AA—inherting the atypical allele from both parents.</td>
</tr>
<tr>
<td>Heterozygous</td>
<td>Two different alleles. Using pseudocholinesterase deficiency as an example, an individual heterozygous for the atypical variant may be considered UA—inherting 1 normal copy and 1 variant copy.</td>
</tr>
<tr>
<td>Genotype</td>
<td>Appearance of the genetic material</td>
</tr>
<tr>
<td>Phenotype</td>
<td>Outward appearance, more commonly described as “what the person looks like.”</td>
</tr>
<tr>
<td>Autosomal dominant inheritance</td>
<td>Mode of inheritance that requires only 1 abnormal gene in order to have the condition (eg, Huntington disease). It is common in this mode of inheritance to see 1 or both parents affected. The chance of inheriting the disease is 50% if 1 parent is heterozygous for the mutant gene and the other is homozygous for the normal gene.</td>
</tr>
<tr>
<td>Autosomal recessive inheritance</td>
<td>Mode of inheritance that requires both copies of the gene to be abnormal in order to have the condition; one abnormal gene from each parent is inherited (eg, cystic fibrosis). Typically the parents are considered carriers and are unaffected. If both parents are heterozygotes for the abnormal gene, there is a 25% chance of having an affected offspring, a 50% chance of a carrier offspring, and 25% chance of an offspring with normal genes. Although pseudocholinesterase deficiency is traditionally considered autosomal recessive, the individual can inherit numerous variants of the gene, including the wild type, and still be considered to have the condition.</td>
</tr>
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</table>
Whittaker, reported the ability to use sodium fluoride as an inhibitor in the testing process. Of the individuals tested, some with normal dibucaine numbers were found to have low fluoride numbers and some with intermediate dibucaine numbers had even lower fluoride numbers. This research led to the discovery of the fluoride-resistant gene. A normal fluoride number is described as 55 to 65. Homozygotes for this gene would be expected to have a moderate sensitivity to succinylcholine.

- **Silent Variant.** The silent variant, although rare and complex, results in the relative lack of pseudocholinesterase activity. Reports of individuals who, when tested, had no pseudocholinesterase activity arose as early as the 1960s. An individual homozygous for the silent variant, when exposed to succinylcholine or mivacurium, will have profound paralysis and apnea, relying solely on alternate pathways for elimination of the drug. Frequencies vary by population, with 1 in 100,000 individuals of European or American descent being homozygous for the variant, whereas individuals in the Vysya community of India have a frequency rate of 4%.1

- **K Variant.** The K variant is the most common variant of the \( \text{BChE} \) gene and is the final variant to be discussed. The K variant is responsible for a 30% reduction in enzyme activity, which is of minimal clinical significance if occurring alone. However, in 89% of cases in which the atypical variant was identified, the K variant also existed, making the K variant more clinically significant.8 Additionally, the combination of the K variant and an acquired deficiency may lower enzyme activity to levels at which one would see a clinical effect. The recognition of the K variant is difficult using traditional biochemical assays, and for this reason many individuals previously tested may have been misclassified in the past. Accurate identification of this variant requires DNA sequencing.8

### Table 2. Common Genetic Variants Found in Pseudocholinesterase Deficiency and Anesthetic Implications After Succinylcholine

<table>
<thead>
<tr>
<th>Variant</th>
<th>Frequency(^a)</th>
<th>Activity</th>
<th>Sensitivity</th>
<th>Estimated paralysis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wild type (U)</td>
<td>Reported to be as high as 98% of the general population</td>
<td>Normal enzyme activity</td>
<td>Normal response to succinylcholine (UU)</td>
<td>Normal: ~5 min</td>
</tr>
<tr>
<td>Atypical (A)/dibucaine resistant</td>
<td>Homozygote: 1/3,000 - 1/10,000 (0.03%-0.01%)</td>
<td>Homozygote: activity decreased by 70%</td>
<td>Homozygote (AA): very sensitive</td>
<td>≥2 h</td>
</tr>
<tr>
<td></td>
<td>Heterozygote: up to 1/25 (4%)</td>
<td></td>
<td>Heterozygote (UA): occasional prolonged apnea</td>
<td></td>
</tr>
<tr>
<td>Fluoride resistant (F)</td>
<td>Homozygote: 1/150,000 (0.0007%); heterozygous condition is more common but less clinically significant</td>
<td>Activity decreased by 60%</td>
<td>Homozygotes (FF): moderately sensitive</td>
<td>1-2 h</td>
</tr>
<tr>
<td>Silent (S)</td>
<td>Homozygous: 1/10,000 - 8/100,000 (0.01%-0.008%); heterozygous condition is more common but less clinically significant</td>
<td>No activity</td>
<td>Homozygotes (SS): extremely sensitive</td>
<td>3-4 h or more</td>
</tr>
<tr>
<td>Kalow (K)</td>
<td>Homozygous: 1/65 (1.5%); often associated with other variants</td>
<td>Activity decreased by 30%</td>
<td>Homozygotes (KK): → mildly sensitive</td>
<td>&lt;1 h</td>
</tr>
</tbody>
</table>

\(^a\) Frequency distribution for the general population; frequencies and existence of different variants vary among different populations. 
\(^b\) Variants can exist by themselves or in combination with other variants, making the response to succinylcholine (or mivacurium) more variable than as highlighted in the table. This table illustrates only individuals homozygous for 1 specific allelic variant.

### Testing for Pseudocholinesterase Deficiency

- **Biochemical Assays.** Serum cholinesterase activity can determine if there is a quantitative defect in enzyme function. “Normal” is described as 3,200 to 6,600 IU/L. This number varies by different laboratory standards and is subject to much interindividual variability. Dibucaine and fluoride are the most common inhibition tests. However, there are many other inhibition tests described in the literature, including chloride, RO2-0683 (one of the inhibitors used in testing), urea, and succinylcholine. Although these tests are beneficial in determining whether a deficiency exists, they are not always accurate in distinguishing the individual genotype, which requires genetic testing.8

- **Molecular Testing.** Polymerase chain reaction (PCR) is a process used to replicate specific pieces of DNA. The goal of this test is to identify where and what mutations or variants exist on a particular piece of DNA. With this technology qualitative and quantitative variants of the \( \text{BChE} \) gene are able to be identified, and correct genotyping can occur.10 Unfortunately, this technology is available only for research purposes at this time.
Acquired Conditions Affecting Pseudocholinesterase Activity

- **Liver Disease.** Pseudocholinesterase is primarily synthesized in the liver. When liver function is impaired, pseudocholinesterase synthesis is also impaired. Decreased serum activities have been shown in many liver diseases, such as cirrhosis, end-stage liver disease, hepatitis, and liver abscesses. In patients with end-stage liver disease, normal serum pseudocholinesterase levels were again seen after liver transplant, with the transplanted liver assuming the role of production immediately.\(^{11,12}\) Additionally, serum cholinesterase activity may drop 30% to 50% in acute hepatitis, with a 50% decrease in cirrhosis and chronic malignancies being perhaps among some of the most substantial decreases of the acquired conditions.\(^ {13}\) As the half-life of serum cholinesterase is approximately 10 to 14 days, it is considered an unreliable source for tracking liver disease.\(^ {13,14}\)

- **Renal Disease.** Like liver disease, the literature establishes a clear connection between renal disease and decreased levels of pseudocholinesterase, although the reasons are not as clear-cut.\(^ {15,16}\) Serum cholinesterase activity was found to be up to 2 standard deviations below normal in 60% of individuals with renal failure who were studied.\(^ {15}\) Further work shows that this decrease cannot be attributed to the use of dialysis, as previously suspected. Those undergoing renal transplantation experienced an initial drop in pseudocholinesterase activity, with subsequent normalization approximately 15 days after transplant, roughly the same half-life as the enzyme.\(^ {17,18}\)

- **Malnutrition.** Malnutrition is closely linked with changes in serum albumin and pseudocholinesterase, likely due to changes in hepatic synthesis of the proteins and enzymes associated with this disease. In one particular study, 83% of children who were clinically considered malnourished had serum pseudocholinesterase levels below the lower threshold of normal. Degenerative changes of the liver were shown in all 43 samples taken in this study.\(^ {19}\) Although this may be an extreme illustration, as providers we must be aware of these changes because individuals with varying degrees of malnourishment may present to us—possibly anorexic, elderly, homeless, or alcoholic individuals. In addition, this information may be substantial for individuals who volunteer their time and efforts in underprivileged areas of the world. Again, clear case reports exist linking prolonged paralysis and apnea after succinylcholine administration in people with malnourishment.

- **Pregnancy.** Reductions in pseudocholinesterase activity are said to begin approximately in the 10th week of pregnancy, with further decreases postpartum before normalizing between 10 days and 6 weeks postpartum.\(^ {20}\) Activity is said to be reduced 24% during pregnancy, 25% at 1 day postpartum, and 33% at 3 days postpartum. These reductions themselves are not clinically significant, as it has been suggested that activity needs to be 50% or less of average activity for individuals to show sensitivity to succinylcholine.\(^ {14}\) More importantly, 60% of individuals with HELLP syndrome (hemolysis, elevated liver enzymes, and low platelet count occurring in association with preeclampsia) at the time of the study showed pseudocholinesterase activity below the limit of normal. The authors speculate this reduction could in fact be clinically significant if exposed to succinylcholine, and they postulated that this effect is due to liver damage seen in HELLP syndrome.\(^ {21}\) It is important to use caution when choosing an anesthetic plan for the pregnant patient because normal reductions combined with a genetic variant could prove clinically significant if exposed to succinylcholine or an ester local-like chloroprocaine.

- **Malignancy.** Decreased levels of pseudocholinesterase have been associated with malignancy and carcinoma, with decreases closely related to the site of primary lesion and degree of spread.\(^ {22}\) Hepatic carcinomas demonstrate the greatest degree of reductions, followed by lung, gastrointestinal, and genitourinary malignancies, with breast cancer having a smaller effect on enzyme levels. Evidence of abnormal cholinesterase gene expression in many tumor types exists, with great variation among different tumors.\(^ {11,23}\) Consider this information when deciding which muscle relaxant to use when, for example, securing an airway on an individual with stage IV lung cancer.

- **Burns.** The size and severity of a burn closely correlates with reductions in pseudocholinesterase activity.\(^ {24}\) Lowest levels have been found 5 to 6 days after a burn, with levels sometimes being depressed 80% or greater, and in severely burned patients levels remained low up to 4 months after initial injury. It is hypothesized that the initial drop in activity is due to dilution effect and transcapillary losses, whereas prolonged decreases are due to hepatic depression of synthesis or release and/or the presence of inhibiting substances in the plasma.\(^ {24}\) The importance of this information is questionable, as succinylcholine is essentially contraindicated in the burn patient because of the risk of hyperkalemia.

- **Cardiopulmonary Bypass.** A recent study indicates reductions in pseudocholinesterase activity up to 37% with the initiation of cardiopulmonary bypass, with values remaining low after the termination of bypass.\(^ {25}\) This reduction was said to be unrelated to anesthetic technique, including the use of heparin and aprotinin, but rather related to hemodilution.\(^ {25}\) Like other acquired conditions, the reduction seen here may be of little clinical significance, but in combination with a genetic variant, relevant condition, or presence of a particular drug, the reduction may be a contributing factor to prolonged apnea and paralysis after succinylcholine administration.

- **Leprosy.** Increased incidences of pseudocholinesterase deficiency have been found in populations with leprosy compared with control groups.\(^ {26}\) The most striking inci-
Pseudo cholinesterase deficiency is found in individuals with lepromatous leprosy, in which the individual lacks immune resistance to the *Mycobacterium leprae* organism. Researchers have speculated that there may be a possible genetic link between pseudocholinesterase deficiency and susceptibility to leprosy.

### Drug Influences on Pseudocholinesterase

Refer to Table 3 for a comprehensive list of drugs that influence pseudocholinesterase activities.

- **Anesthesia-Related Drugs: Anticholinesterases and Pancuronium.** It is very common, in any given day, to use an anticholinesterase to reverse a nondepolarizing muscle relaxant. However, their role in reversing succinylcholine-induced apnea is controversial and has met with mixed results. Anticholinesterases, by definition, inhibit cholinesterases, thereby inhibiting the breakdown of succinylcholine. The literature is abundant with case reports of prolonged apnea and paralysis, incomplete antagonism, and potentiation of blockade when succinylcholine is combined with anticholinesterases, including neostigmine, pyridostigmine, edrophonium, and physostigmine. It has been further suggested that large doses of neostigmine can cause paradoxical blockade.

Pancuronium is a nondepolarizing muscle relaxant, which possesses some anticholinesterase activity. Early publications show a depression of serum cholinesterase of 40% up to 45 minutes after injection of pancuronium. More recently, individuals with the normal genotype were given standard clinical doses of pancuronium for elective surgical procedures. Nearly all individuals showed a statistically significant decrease in pseudocholinesterase activity, but all levels were considered to be in the normal range. Again, the clinical significance of these findings may be questioned, because pancuronium is used for procedures in which a longer period of relaxation is required and reductions in enzyme activity do not reach a clinically significant level.

Although many of the drugs mentioned in Table 3 do not cause clinically relevant reductions in pseudocholinesterase activity, they are important to recognize. Whereas drugs, certain genetic variants, or acquired conditions alone may not be enough to see prolonged paral-

<table>
<thead>
<tr>
<th>Drug</th>
<th>Influence</th>
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<tbody>
<tr>
<td>Echothioephate eyedrops</td>
<td>Succinylcholine-induced apnea was reached within days of using the eyedrops. Should be discontinued several weeks before use of succinylcholine is considered.</td>
</tr>
<tr>
<td>Organophosphate insecticides</td>
<td>Many different “brands.” Readily absorbed through bronchi, intact skin, and GI tract. In acute poisoning, pseudocholinesterase activity is 30%-50% of normal. Numerous case reports in literature of prolonged apnea and paralysis after exposure to succinylcholine and organophosphate insecticides.</td>
</tr>
<tr>
<td>Cyclophosphamide</td>
<td>Dose-dependent reduction in pseudocholinesterase activity</td>
</tr>
<tr>
<td>Neostigmine, pyrnostigmine, pyridostigmine, edrophonium</td>
<td>Case reports of prolonged apnea and paralysis, incomplete antagonism, and potentiation of blockade when succinylcholine is combined with anticholinesterases including neostigmine, pyridostigmine, edrophonium, and physostigmine. Considered controversial for reversal of succinylcholine-induced apnea and paralysis.</td>
</tr>
<tr>
<td>Tacrine</td>
<td>Used for Alzheimer disease therapy. Was used in the 1960s to increase duration of paralysis by up to 11 minutes when used with succinylcholine.</td>
</tr>
<tr>
<td>Cocaine</td>
<td>Ester local anesthetic. Up to 50% of cocaine is reported to be metabolized by pseudocholinesterase. Decreased plasma cholinesterase activity is associated with increased risk of life-threatening cocaine toxicity.</td>
</tr>
<tr>
<td>Oral contraceptives</td>
<td>Enzyme activity reduced approximately 20%; likely related to steroid-induced depression of the liver’s ability to synthesize or secrete into circulation.</td>
</tr>
<tr>
<td>Monoamine oxidase inhibitor: phenelzine</td>
<td>Case reports of prolonged apnea and paralysis after ECT when individuals were receiving phenelzine. Pseudocholinesterase levels normalized after withdrawal from the drug.</td>
</tr>
<tr>
<td>Pancuronium</td>
<td>Enzyme activity reduced up to 40% 45 minutes after a dose. Significant reductions observed but still considered within normal range.</td>
</tr>
<tr>
<td>Bambutrol (oral bronchodilator)</td>
<td>Plasma cholinesterase activity reduced a median of 90%, increasing duration of action of mivacurium 3- to 4-fold compared with controls.</td>
</tr>
<tr>
<td>Aspirin</td>
<td>Up to 90% hydrolyzed by cholinesterase. Individuals with pseudocholinesterase deficiency will hydrolyze aspirin more slowly, possibly prolonging its effects.</td>
</tr>
<tr>
<td>Metoclopramide (Reglan)</td>
<td>Administration may prolong block 2-3 minutes; however, if succinylcholine is administered after metoclopramide, especially in individuals who at baseline have low activity or deficiency, a clinically significant prolonged block may occur.</td>
</tr>
</tbody>
</table>

**Table 3. Drugs and Pseudocholinesterase**

GI indicates gastrointestinal; ECT, electroconvulsive therapy.
ysis and apnea following succinylcholine administration, the combination of them could result in a clinically significant scenario.

**Treatment of Pseudocholinesterase Deficiency**

There is no cure for pseudocholinesterase deficiency. However, there are interventions in the literature that serve to, in some cases, speed the onset of recovery once an individual is exposed to succinylcholine or mivacurium. Administration of whole blood, fresh frozen plasma, and human serum cholinesterase are some of the interventions discussed in the literature. The standard of care is widely advocated as letting the individual recover spontaneously.

Administration of whole blood has been successfully described in the literature as reversing prolonged apnea and paralysis after the administration of succinylcholine in individuals with pseudocholinesterase deficiency. Fresh frozen plasma has been met with some controversy, because it shows acceleration of recovery in some individuals and no effect in others. Even though blood and blood product transfusion is generally considered safe, some question why we would unnecessarily expose an individual to a transfusion when it has been clearly established that spontaneous recovery occurs without any undo effects.

Another treatment described in the literature is the use of human serum cholinesterase, which like other interventions, has proved to shorten the recovery time. The product is considered safe, and the disease transmission risk is said to be comparable to that of human albumin. This may be a viable option, but a review of the literature shows that this product is expensive and is not available in the United States.

Although multiple interventions have been described in the literature to speed the onset of recovery from prolonged paralysis and apnea in the pseudocholinesterase-deficient patient, these interventions may pose an unnecessary risk. All individuals will eventually recover spontaneously from prolonged paralysis and apnea, with time to recovery depending on both genetic and acquired factors. It is widely advocated that the best and safest “treatment” is to let the individual recover spontaneously. Once it is suspected that an individual may be pseudocholinesterase deficient, appropriate laboratory work should be drawn and sent off to identify if the individual is pseudocholinesterase deficient. At the same time, it is important to communicate with the patient to help alleviate any distress he or she might be feeling, and then to sedate the patient in the postanesthesia care unit (PACU) or ICU setting until patient recovery is sufficient to meet extubation criteria. Furthermore, the literature describes the need for family members to be tested and all those with pseudocholinesterase deficiency to carry a medical identification (ID) card or bracelet.

**Summary**

Pseudocholinesterase deficiency is a genetic or acquired condition in which the metabolism of succinylcholine, mivacurium, and ester local anesthetics could potentially be impaired. The most recognizable feature of this deficiency is prolonged periods of apnea and paralysis following clinical doses of succinylcholine or mivacurium. Healthcare providers must have an understanding of the genetics of the deficiency as well as conditions and drugs that can alter enzyme activity in order to effectively communicate with patients, their families, and colleagues about this disorder. Helpful websites on genetics are listed in Table 4.

Several different genetic variants exist, each with their own anesthetic implications. Growing genetic research points to different variants coexisting, further prolonging apnea and paralysis after succinylcholine administration. In particular, the K variant alone produces a decrease in enzyme activity of only 30% but when paired with other variants, acquired conditions, or drugs, it can result in reductions of enzyme activity to clinically significant levels. The importance of a thorough preoperative evaluation, including personal and family history, cannot be overemphasized in the detection of a possible deficiency, whether genetic or acquired.

All anesthesia providers should employ the use of a peripheral nerve stimulator when using any muscle relaxant, including succinylcholine. Recovery of twitches should be ensured before administering a nondepolarizing muscle relaxant. If the patient shows no response to train-of-4 stimulation after 15 minutes, and after appropriate troubleshooting, the provider should suspect a pseudocholinesterase deficiency. Appropriate, facility-dependent laboratory work should be immediately sent off. These tests
may include serum pseudocholinesterase activity and dibucaine and fluoride inhibition tests. Efforts should be made to inform the patient of the events in an effort to alleviate stress. The patient should be sedated and ventilated in the PACU or ICU setting until extubation criteria is met. If this situation is encountered in an outpatient surgery center, the patient should be transferred to a facility for longer-term ventilation and sedation.

The most widely advocated method of “treatment” of pseudocholinesterase deficiency is spontaneous recovery. It is the job of the healthcare provider to communicate to the patient and family members information about the condition, implications, inheritance, and testing. Individuals with inherited deficiency should carry a medical knowledge base regarding all aspects of this condition will help healthcare providers to provide accurate information to all healthcare providers. Again, a sound knowledge base regarding all aspects of this condition will help healthcare providers to provide accurate information to patients, families, and colleagues.

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AUTHORS

Flanna K. Soliday, CRNA, MSN, is a staff nurse anesthetist at Forbes Regional Campus of the Western Pennsylvania Hospital, Monroeville, Pennsylvania. At the time this article was written, she was a student at the University of Pittsburgh, Nurse Anesthesia Program, Pittsburgh, Pennsylvania. Email: jifter@comcast.net.

Yvette P. Conley, PhD, is an associate professor of nursing and human genetics at the University of Pittsburgh. Email: yconley@pitt.edu.

Richard Henker, CRNA, PhD, is an associate professor and vice chair of the Acute/Tertiary Care Department at the University of Pittsburgh School of Nursing. Email: rhe001@pitt.edu.