Stiff-person syndrome (SPS) is a neurologic disorder characterized by painful involuntary episodes of severe muscle rigidity affecting the axial muscles and extremities. Although the etiology of SPS is unknown, it is suspected to involve the synthesis of \( \gamma \)-aminobutyric acid (GABA). Symptoms of SPS are precipitated by sudden unexpected movements, noises, and stress. Additionally, SPS has been linked with various autoimmune disorders, including diabetes mellitus, thyroid disease, pernicious anemia, and certain cancers. Because of the effect of SPS and SPS medications, inhalational agents and neuromuscular blockers have the potential to cause prolonged hypotonia following anesthesia, resulting in respiratory failure despite full reversal of neuromuscular blockade. In documented case reports, the outcomes of using general anesthesia with inhalational agents and neuromuscular blockers in patients with SPS varied. This case report highlights the anesthetic management of a 56-year-old woman with diagnosed SPS undergoing a hemicolectomy for a colon mass using total intravenous anesthesia.

Keywords: Moersch-Woltman syndrome, stiff-man syndrome, stiff-person syndrome, stiff-limb syndrome, total intravenous anesthesia.
affects the central nervous system. In some variants of SPS it is hypothesized the syndrome manifests from the body’s immune response to eliminate cancer cells. Because of the similarities between the tumor antigen and central nervous system (CNS) antigens, high levels of GAD antibodies attack the CNS. High levels of GAD antibodies are found in a variety of other neurologic disorders such as cerebellar ataxia, epilepsy, idiopathic limbic encephalitis, and myasthenia gravis. However, Newton et al mentioned that antibodies to GAD alone do not indicate a definitive diagnosis of SPS. Patients can test GAD negative but still exhibit symptoms of SPS.

The clinical manifestations of SPS can occur randomly or be triggered by emotional stress and noxious stimuli such as noise and sudden touch. The diagnosis is made in the presence of episodes of muscle spasms and stiffness of axial muscles slowly progressing to extremities and deformity of the spinal column (severe lordotic curve). Patients often present with a wide stance and an unsteady gait. The gait resembles waddling and has been referred to as tin man like. Patients with SPS also have a hunched-over posture and abnormal eye movements, experience frequent falls, and are often bedridden. Moersch and Woltman described that SPS may be mistaken for chronic tetanus, but it can also be misdiagnosed as anxiety disorder, multiple sclerosis, Parkinson disease, and somatic disorders. Misdiagnosis is common and a correct diagnosis takes an average of 6.2 years from the onset of SPS symptoms.

Presently no cure for SPS exists, but the primary treatment is to enhance GABA receptors by targeting GABA with baclofen therapy, a GABAergic agonist, and targeting GABA with benzodiazepine agents such as diazepam. Patients requiring higher doses of baclofen may be treated via intrathecal baclofen pumps. Spinal cord stimulation may relieve pain associated with spasms. Intravenous (IV) opiates can relieve severe pain episodes and botulinum toxin A injections into the muscles can mitigate severe muscle rigidity. Intravenous immunotherapy such as IV immunoglobulin (IVIG) can modulate the immune response. Corticosteroids and plasmapheresis are also recommended to suppress the antibody response and remove the antibodies respectively. Furthermore, in a recent case report by Vicente-Valor et al, an alternative treatment with cannabis derivatives, tetrahydrocannabinol and cannabidiol oromucosal spray, was successfully used to treat the spasticity symptoms related to SPS.

Case Summary
A 56-year-old African American woman with a diagnosis of SPS was scheduled for a left hemicolectomy for removal of a colon mass. She presented with a body mass index of 16.9 kg/m² (height of 1.753 m and weight of 51.71 kg) after a recent weight loss of 15.75 kg. Her SPS was diagnosed 17 years before her surgery and initially presented with severe abdominal spasms and gagging episodes. At the time of diagnosis, GAD antibodies were elevated to more than 600 U/mL (normal, < 1 U/mL). Her reported activity was limited, as she was wheelchair bound with a severe lordotic curve, but she remained fairly independent. Her symptoms were managed pharmacologically with diazepam, 10 mg 3 times a day; baclofen, 20 mg 4 times a day; tizanidine, 2 mg 3 times a day; and IVIG infusions for flare-ups as needed. Her last infusion was 1 month before surgery.

The patient’s medical history also included asthma, osteoporosis, hyperlipidemia, gastroesophageal reflux disease (GERD), uncontrolled type 1 diabetes with hyperglycemic episodes (last glycated hemoglobin, or HbA1C, was 11.3%), hypothyroidism, pernicious anemia, controlled seizures, and rheumatoid arthritis.

An echocardiogram completed 1 month before the scheduled hemicolectomy showed an ejection fraction of 60% to 65% with normal diastolic function, but left ventricular hypertrophy was apparent. An electrocardiogram (ECG) showed a prolonged QT interval with a T-wave abnormality in the inferior leads, a new finding from the prior ECG 2 years before the scheduled hemicolectomy. On preoperative interview she denied any recent chest pain, shortness of breath, palpitations, dizziness, new symptoms of spasticity, gagging movements, or abdominal cramping. Specifically, she denied spasticity since a prior admission 5 weeks earlier when she received IVIG. She also conveyed that she commonly has a spastic episode when she discontinues her SPS medications. Before the scheduled hemicolectomy the patient had several episodes of severe hypoglycemia, which required hospitalization and treatment with 50% dextrose, with the last admission being 3 weeks before the scheduled procedure. In the preoperative clinic she was advised to continue her SPS medications, but despite this recommendation she withheld her SPS medications the day of surgery.

The morning of surgery, the patient was asymptotically hypoglycemic with a blood glucose level of 70 mg/dL. Her regimen consisted of metformin at 1 g/d and an insulin glargine injection (Lantus) of 20 U nightly. Her last dose of insulin was the evening before surgery. She was treated with 1.5 ampules of 50% dextrose, which increased her glucose level to 160 mg/dL preoperatively. The patient’s surgical history included cholecystectomy, hernia repair, and multiple colonoscopies managed with inhalational anesthesia and monitored anesthesia care (MAC) techniques that yielded no adverse outcomes. Airway assessment showed Mallampati classification 2, thyromental distance greater than 3 fingerbreadths, mouth opening greater than 3 cm, and limited neck extension.

After preoperative assessment was reviewed and informed consent was obtained, the patient was given midazolam, 2 mg IV, before proceeding into the operat-
ing room. The operating room was quiet and warmed before the patient's arrival. Standard ASA monitors were applied, and the patient was given 100% oxygen via face mask. Cricoid pressure was applied to the patient throughout induction and intubation given her history of uncontrolled GERD. Induction agents consisted of a titration of fentanyl, 100 μg; lidocaine, 100 mg; and propofol, 200 mg, IV. Tracheal intubation with a 7.0-mm oral standard endotracheal tube was easily achieved without the use of a muscle relaxant via a size 3 Macintosh laryngoscope blade providing a grade 1 view. A 20-gauge arterial line was placed in the right radial artery. An 18-gauge peripheral IV was placed to the left forearm. A peripheral nerve stimulator was applied over the ulnar nerve. The patient’s severe lordotic curve was padded along with other pressure points. General anesthesia was maintained with an oxygen and air mixture, propofol infusion was titrated between 120 and 200 μg/kg/min, and a remifentanil infusion was titrated between 0.1 and 0.25 μg/kg/min to achieve a bispectral index of 40 to 60.

The initial anesthesia plan was to use a paralytic agent throughout the case as needed to facilitate adequate surgical conditions. Vecuronium, 2 mg, was chosen for its intermediate action and predictability. This agent provided adequate muscle relaxation for approximately 20 minutes to achieve a train-of-four (TOF) of 2/4 for a midline incision of the abdominal muscles and surgical exposure. The patient returned to a TOF of 4/4, and after discussion with the surgeon who believed that operative conditions were adequate, the decision was made to forgo additional paralytics. Propofol, administered as a 100-mg bolus in addition to the propofol and remifentanil infusions, provided adequate surgical conditions for closure of the abdominal muscles. In addition to 2 L of balanced electrolytes (Normosol-R), 500 mL of 5% dextrose in water was infused at 170 mL/h throughout the case (3 hours), and the blood glucose level was checked every hour to monitor for hypoglycemia. The patient's glucose levels ranged from 70 to 160 mg/dL throughout the perioperative period. The total blood loss was 250 mL and urine output was 300 mL. Ondansetron, 4 mg, was given IV before emergence.

Before extubation the patient had a TOF of 4/4 with sustained tetany for 5 seconds showing no fade, a tidal volume of 350 to 400 mL (6-8 mL/kg), and a sustained head lift greater than 3 seconds with eye opening. The patient was not administered neuromuscular blockade reversal because sustained tetany showed no fade and she did not exhibit signs of muscle weakness. The patient was extubated to 100% oxygen via face mask 10 minutes after the remifentanil and propofol infusions were stopped. Hydromorphone, 6 mg IV, was titrated for pain management, and promethazine (Phenergan), 18.7 mg IV, was titrated for control of nausea.

She was transferred to the surgical intensive care unit (SICU) on room air for close postoperative monitoring without any complications. Vital signs on SICU admission were blood pressure of 123/72 mm Hg, pulse of 90/min, temperature of 36.6°C, respirations of 18/min; pulse oximetry was 98%, and the glucose level was 150 mg/dL.

Postoperatively the patient was managed by the colorectal surgery service with consultation to both neurology and internal medicine. She received morphine, 5 mg IV, as needed for pain and was restarted on her home medication regimen for SPS, including diazepam, 10 mg 3 times a day; baclofen, 20 mg 4 times daily; tizanidine, 2 mg 3 times a day; and hyoscyamine, 0.125 mg every 4 hours as needed for cramping. Her antiepileptic regimen also was restarted, which included levetiracetam (Keppra), 500 mg twice a day, and phenytoin, 200 mg twice daily (dosage decreased from 300 mg twice a day as her serum phenytoin level was elevated). She was also continued on a regimen of 5% dextrose with half normal saline with 20 mEq of potassium chloride at 100 mL/h because of concerns for recurrent hypoglycemia and mild hypokalemia.

On postoperative day 2 the patient transitioned from clear liquids to a regular diabetic diet, at which time she moved to the general surgical unit. She did have a decrease in her hemoglobin level to 7.5 g/dL on postoperative day 2, which required transfusion of 1 U of packed red blood cells with adequate response. Internal medicine was consulted for diabetes management and recommended that after discharge she be continued on a regimen of insulin glargine, 10 U at bedtime, and metformin, 1 g twice daily. She was able to tolerate a regular diet and did not have any new symptoms of spasticity or rigidity.

She remained stable and was discharged home without any incident on postoperative day 4. The patient was followed up in the colorectal surgery clinic 3 weeks after the procedure and did not have any complaints at that visit.

**Discussion**

In our case we successfully used propofol and remifentanil infusions with minimal paralytics, resulting in no hypotonia at the end of the case. The patient did not report any adverse events with her previous surgery, for which an inhalational agent was used; however, the records were unavailable, and the patient did not know the dates of the surgeries. Since the time of her previous surgeries, it was unclear whether her symptoms had progressed and SPS treatments could have been increased substantially. Although regional anesthesia is an appropriate option, we elected not to use this method because of the patient's severe spinal deformity, anxiety, failure to take her SPS medications the day of surgery, and the risk of initiating a spasm while placing the epidural/spinal anesthetic.

In a search of the medical literature, only a small number of published case studies were available. The outcomes of using general anesthesia with inhalational agents
in patients with SPS are varied (Table 1). Of the 17 cases we reviewed related to SPS, 7 cases used inhalational anesthesia, 2 cases used regional anesthesia combined with MAC, and 2 cases used a sole MAC technique (Table 2). Five case reports we found used a total intravenous anesthesia (TIVA) technique (Table 3).

- **Preoperative Considerations.** Preoperatively, patients should continue their immunotherapy, benzodiazepine, and baclofen to optimize the suppression of their SPS symptoms. Baclofen, if stopped abruptly, can cause withdrawal symptoms and may require oral baclofen. Baclofen withdrawal symptoms mimic alcohol withdrawal and neuroleptic malignant syndrome. The symptoms associated with withdrawal include, but are not limited to, seizures, hallucinations, sweating, confusion, labile heart rate and blood pressure, fever, and increased spasticity. Although there is no association with any cardiorespiratory disorders, it is recommended to perform a pulmonary

### Table 1. Case Studies of Stiff-Person Syndrome Receiving Either General Anesthesia or Regional Anesthesia Combined With General Anesthesia

<table>
<thead>
<tr>
<th>Author/patient sex/age, y</th>
<th>SPS medications and dosage</th>
<th>Surgical procedure</th>
<th>Drugs administered</th>
<th>Postoperative outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bouw19/F/62</td>
<td>Diazepam, 7.5 mg twice a day; baclofen, 12.5 mg twice a day; prednisone, 20 mg/d</td>
<td>Colon resection</td>
<td>Isoflurane, propofol, sufentanil, atracurium, morphine, neostigmine, glycopyrrolate</td>
<td>Prolong intubation 3 hours into the postoperative period</td>
</tr>
<tr>
<td>Johnson20/F/46</td>
<td>Diazepam 100 mg/d; intrathecal baclofen pumpa</td>
<td>Baclofen pump</td>
<td>Propofol, succinylcholine, desflurane</td>
<td>Weakness 2 days postoperatively</td>
</tr>
<tr>
<td></td>
<td>Baclofen pump repair 1</td>
<td></td>
<td>Isoflurane, nitrous oxide, sufentanil, thiopental, vecuronium, neostigmine, glycopyrrolate</td>
<td>Muscle weakness and hypotonia present; patient was intubated overnight in ICU</td>
</tr>
<tr>
<td></td>
<td>Baclofen pump repair 2</td>
<td></td>
<td>Midazolam, halothane, isoflurane, nitrous oxide</td>
<td>No exacerbation of symptoms present</td>
</tr>
<tr>
<td>Obara18/F/40</td>
<td>Not given</td>
<td>Thymectomy</td>
<td>Fentanyl, thiopental, vecuronium, isoflurane, nitrous oxide</td>
<td>No exacerbation of symptoms present</td>
</tr>
<tr>
<td></td>
<td>Appendectomy</td>
<td></td>
<td>Diazepam, fentanyl, thiopental, vecuronium, isoflurane, nitrous oxide</td>
<td>No exacerbation of symptoms present</td>
</tr>
<tr>
<td>Qin21/M/58</td>
<td>Diazepam, 5 mg twice a day; baclofen, 10 mg 3 times a day</td>
<td>Thymectomy</td>
<td>Midazolam, propofol, remifentanil, rocuronium, isoflurane, nitrous oxide</td>
<td>No exacerbation of symptoms present</td>
</tr>
<tr>
<td>Yamamoto22/M/76</td>
<td>Diazepam, 5 mg/d; baclofen, 30 mg/d; clonazepam, 1 mg/d</td>
<td>Thymectomy</td>
<td>Sevoflurane, fentanyl, propofol, ropivacaine, 0.25% infusion 24 hours postoperatively</td>
<td>No exacerbation of symptoms present</td>
</tr>
</tbody>
</table>

Abbreviations: ICU, intensive care unit; SPS, stiff-person syndrome.

aThe dose was not provided in the case report.

(From Ferrandis et al and Sidransky, Tran, Kaye24)
function test to determine if respiratory insufficiencies are present and to aid in determining a postoperative plan for possible respiratory support. Laboratory results, including electrolytes, coagulation panel, and GAD antibody levels, should be checked before surgery. Levels of GAD antibodies may indicate the severity of the disease and predict response to medications and immunotherapy. Coagulation laboratory findings will aid in deciding if spinal or epidural anesthesia is an option for the anesthetic plan. It is optimal to schedule IVIG therapy close to the surgery date to decrease the levels of GAD antibodies, thus decreasing the chance of a spasm occurring.

**Intraoperative Considerations.** The operating room should be warmed and quiet. Any loud noises should be diminished as much as possible because it can elicit a spastic episode. In the case studies we reviewed, patients with SPS are not characterized by having difficult airways. As a result of muscle rigidity, patient positioning can present a challenge from difficulty flexing and extending muscles. The patient's lordotic curve and other

<table>
<thead>
<tr>
<th>Method of anesthesia</th>
<th>Author/ patient sex/age, y</th>
<th>SPS medications and dosage</th>
<th>Surgical procedure</th>
<th>Drugs administered</th>
<th>Postoperative outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Regional anesthesia (paravertebral block and MAC)</td>
<td>Elkassabany11/M/65</td>
<td>Intrathecal baclofen pump&lt;sup&gt;a&lt;/sup&gt;</td>
<td>Inguinal hernia</td>
<td>Bupivacaine 0.5% Midazolam Fentanyl Propofol</td>
<td>Patient reported improvement of spasticity symptoms during procedure and 1 hour postoperatively</td>
</tr>
<tr>
<td>Regional anesthesia (spinal/ epidural)</td>
<td>Shanthanna23/F/55</td>
<td>Baclofen, clonazepam, gabapentin&lt;sup&gt;a&lt;/sup&gt;</td>
<td>Bilateral amputation of lower extremities</td>
<td>Bupivacaine 0.5% Midazolam Fentanyl</td>
<td>Patient did not experience exacerbation of symptoms; however, anesthesia provider was unable to achieve an appropriate anesthesia level without supplementing with epidural</td>
</tr>
<tr>
<td>MAC</td>
<td>Sidransky24/M/34</td>
<td>Carisoprodol, 250 mg/d; diazepam, 10 mg twice a day; gabapentin, 600 mg 3 times a day; baclofen, 30 mg/d</td>
<td>Permanent catheter placement</td>
<td>Lidocaine Propofol</td>
<td>No exacerbation of symptoms present</td>
</tr>
<tr>
<td></td>
<td>Neubert2/M/55</td>
<td>Not given</td>
<td>Left ilioinguinal nerve block</td>
<td>Midazolam Ketamine</td>
<td>No exacerbation of symptoms present</td>
</tr>
</tbody>
</table>

Table 2. Case Studies of Stiff-Person Syndrome Receiving Regional Anesthesia or Monitored Anesthesia Care

Abbreviations: MAC, monitored anesthesia care; SPS, stiff-person syndrome.

<sup>a</sup>The dose was not provided in the case report.

(From Ferrandis et al<sup>1</sup> and Sidransky, Tran, Kaye<sup>24</sup>)

<table>
<thead>
<tr>
<th>Author/patient sex/age, y</th>
<th>SPS medications and dosage</th>
<th>Surgical procedure</th>
<th>Drugs administered</th>
<th>Postoperative outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ferrandis1/F/44</td>
<td>Diazepam, 50 mg 3 times daily; tizanidine, 4 mg/d</td>
<td>Double heart-valve replacement</td>
<td>Midazolam, diazepam, fentanyl, etomidate, pancuronium, propofol, remifentanil</td>
<td>Hypotonia was not prolonged; however, pain and stiffness with mild contractions in bilateral upper and lower extremities was present 7 hours postoperatively</td>
</tr>
<tr>
<td>Ledowski25/M/74</td>
<td>Clonazepam, quinine, baclofen&lt;sup&gt;a&lt;/sup&gt;</td>
<td>ENT</td>
<td>Remifentanil, propofol</td>
<td>No exacerbation of symptoms present</td>
</tr>
<tr>
<td>Toscano16/F/47</td>
<td>Diazepam&lt;sup&gt;a&lt;/sup&gt;</td>
<td>Incision and drain of breast abscess</td>
<td>Midazolam, fentanyl, propofol, morphine</td>
<td>No exacerbation of symptoms present</td>
</tr>
<tr>
<td>Yagan26/M/46</td>
<td>Diazepam, 15 mg/d; baclofen, 30 mg/d; prednisolone, 20 mg/d</td>
<td>Lumbar vertebral compression fracture repair</td>
<td>Midazolam, lidocaine, propofol, remifentanil</td>
<td>No exacerbation of symptoms present</td>
</tr>
<tr>
<td>Obara18/F/40</td>
<td>Not given</td>
<td>Endoscopic nasal sinus surgery</td>
<td>Propofol, fentanyl, vecuronium,nitrous oxide</td>
<td>No exacerbation of symptoms present</td>
</tr>
</tbody>
</table>

Table 3. Case Studies of Stiff-Person Syndrome Receiving Total Intravenous Anesthesia

Abbreviations: ENT, ear, nose, and throat; SPS, stiff-person syndrome.

<sup>a</sup>The dose was not provided in the case report.

(From Ferrandis et al<sup>1</sup> and Sidransky, Tran, Kaye<sup>24</sup>)
pressure points should be padded.

According to the anesthesia recommendations for patients with SPS made by Shanthanna et al., there are no contraindications to any anesthetic agents or procedures and no particular anesthetic technique has been shown to be safer and more effective than another. When weighing the risks and benefits of general anesthesia vs regional anesthesia, the appropriate anesthetic plan should be chosen based on the provider’s comfort level, location and type of surgery, patient preference, and severity of disease. However, it is hypothesized that inhalational agents exert their effect through GABA inhibition, and they have been reported to potentiate the hypotonia caused by SPS medications, particularly baclofen. In a case unrelated to SPS, a 49-year-old man with a spinal cord injury was scheduled to have a penile implant. The patient was prescribed baclofen, 25 mg 3 times a day, for muscle spasms. On induction, the patient was given fentanyl, thiopental, and atracurium. Anesthesia was maintained with isoflurane and nitrous oxide. At the completion of the surgery, the muscle relaxant was reversed; however, the patient remained intubated because of the presence of muscle hypotonia for 6 hours following the case. It was discovered in the postoperative period that the patient was taking the recommended dose of baclofen and additional doses when muscle spasms were not controlled. Inhalation agents should be avoided if possible. In the event that inhalational agents are used in patients with SPS, hypotonia should be monitored closely before extubation.

In the case report by Bouw et al., a 62-year-old woman was scheduled for resection of a colon mass. Her SPS symptoms were managed with diazepam, 7.5 mg twice a day; baclofen, 12.5 mg twice a day; and prednisone, 20 mg daily. On induction, she was given propofol, sufentanil (Sufenta), and atracurium. Her anesthesia was managed with isoflurane, and despite a TOF of 4/4 and full muscle relaxant reversal, the patient remained intubated 3 hours in the postoperative period because of prolonged hypotonia and respiratory failure. It was hypothesized by the authors that the inhalational agent used furthered the hypotonia by potentiating the effect baclofen has on GABA receptors.

Although the mechanism in unclear, it has also been reported that neuromuscular blockers potentiate the hypotonia caused by SPS medications. In the Johnson and Miller case report, a 46-year-old woman was scheduled for repair of a baclofen pump. Her surgical history included postoperative weakness 2 days following her initial baclofen pump placement. Her SPS symptoms had been managed with diazepam, 100 mg daily, and an intrathecal baclofen pump, in which the dose was not reported. During the follow-up baclofen pump repair surgery, isoflurane and vecuronium were used and resulted in muscle weakness and hypotonia. The patient was intubated and admitted to the intensive care unit (ICU) overnight. When the patient returned to have a baclofen pump repaired 5 months later, inhalation anesthesia without muscle relaxants was used. The patient did not exhibit an exacerbation of symptoms or prolonged hypotonia in the postoperative period. Although it has not been shown that muscle relaxants are prolonged in patients with SPS, it has been recommended that muscle relaxants should be avoided if possible. If muscle relaxants must be used, small doses of short-acting relaxants along with TOF monitoring are recommended. Although no relationship has been found between the depth of muscle relaxation and hypotonia in patients with SPS, it is recommended for muscle relaxants to be reversed. Currently, there is insufficient information on the use of succinylcholine in adults with diagnosed SPS.

Propofol is the recommended drug for the induction and maintenance phase of anesthesia. In previous studies of propofol, it is shown to be beneficial for managing acute muscle spasms associated with SPS and it can also enhance GABA receptors in GABA-deficient patients. In 2 separate studies conducted in rodents, propofol was shown to enhance the GABA receptor activity by shortening the phase of the GABA receptor channel closure and potentiating the GABA receptor-mediated inhibitory postsynaptic currents.

Regional anesthesia is an appropriate option in patients with SPS but can present a challenge to the anesthesia provider. In the Shanthanna case report in which a 55-year-old woman was scheduled for bilateral amputation of the lower extremities, the provider was unable to achieve an appropriate anesthetic level for surgery after a spinal block without supplementing with an epidural. It is noted that the deformity of the spine can cause an unpredictable spinal block, and the rigidity of the muscles can make it difficult to position the patient. Shanthanna mentioned the successful use of regional anesthesia as the safest mode of anesthesia (see Table 2). However, loud noises and emotional stress, such as anxiety can exacerbate spasms. Conscious sedation should be derived in a manner adequate to keep the patient calm before placing the epidural/spinal anesthetic. In the case of a 65-year-old man with SPS undergoing a hernia repair, the anesthesia provider performed regional anesthesia using a paravertebral block, supplementing with a MAC technique. The patient reported improvement of spasticity symptoms during the procedure and 1 hour into the postoperative period.

Postoperative Considerations. Postoperatively the patient should be monitored closely for muscle spasms and weakness that could lead to respiratory failure because of drug interactions between baclofen, diazepam, and anesthetic drugs. All SPS medications should be continued in the postoperative period to prevent muscle rigidity from reoccurring. Patients may require supplementation with benzodiazepines to alleviate postopera-
tive symptoms. In the case study by Ferrandis et al., a 44-year-old woman scheduled for a double heart valve replacement was induced with fentanyl, midazolam, etomidate, and pancuronium. Anesthesia was maintained using TIVA with propofol and remifentanil. Postoperatively she experienced pain in her arms and legs with muscle rigidity in her forearm and lower limbs without spasms. She required a dose of diazepam, 5 mg IV, and morphine, 3 mg IV, in addition to her continuous infusion of midazolam (Versed). Four hours later she exhibited symptoms again and was started on a regimen of diazepam, 50 mg every 8 hours, for pain and stiffness in her legs that eventually subsided. She was discharged from the unit 3 days later without SPS symptoms.\(^1\)

**Conclusion**

Because of the unique effect that SPS has on GABA and the possible interaction of inhalational agents and muscle relaxants with SPS medications, anesthetic management of a patient with SPS can be challenging for the anesthesia provider. In documented case studies TIVA has been used successfully in patients with SPS without causing postoperative hypotonia (see Table 3). The patient's comfort and safety were achieved without any residual weakness before and following emergence and extubation. There were no undesirable symptoms related to SPS noted, suggesting that in our case the use of minimal paralytics, propofol, and opioids can be an appropriate and safe choice to manage patients with SPS.

**REFERENCES**


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**DISCLOSURES**

The authors have declared they have no financial relationships with any commercial interest related to the content of this activity. The authors did not discuss off-label use within the article.