20 Regional Anesthesia in the Presence of Neurologic Disease

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Performing regional anesthesia in patients with preexisting neurologic or neuromuscular disease remains controversial. Numerous anecdotal reports describe the successful use of regional techniques in a variety of neuromuscular disorders including multiple sclerosis, amyotrophic lateral sclerosis (ALS), muscular dystrophies, myotonias, and others.[1] However, a study of significant size to confirm or support the safety of regional anesthesia in these patients continues to remain scarce. Specific guidelines regarding the use of regional techniques in the setting of neurologic disease are difficult to define because of these limitations. Therefore, the goal of this chapter is to review several of the more common neurologic disorders that an anesthesiologist may encounter and outline what information currently exists to help guide the use of regional anesthesia.

Two characteristics of the neuromuscular system that continue to be considered as a contraindication to regional anesthesia include the following: increased intracranial pressure with respect to epidural or spinal anesthesia and evolving or unstable neuromuscular disease. In the background, it is difficult to ignore the literature surrounding cauda equina syndrome in patients who received continuous spinal anesthesia through small-gauge catheters in the past. From this information, higher concentrations of local anesthetics for a longer duration have the potential to be toxic to nerves. More recently, lidocaine more than other local anesthetics has been shown to be neurotoxic in clinically available concentrations, even in short durations. However, there are several disadvantages to general anesthesia that could be detrimental as well. For instance, these patients may have decreased respiratory reserve which may be compromised by even small amounts of narcotics and muscle relaxants. Therefore, judicious use of local anesthetics, especially at lower concentrations, may achieve a middle ground of safety with potentially fewer side effects. This chapter will review some of these disorders and the information available on the use of regional anesthetic techniques in their presence.

General Considerations

Evaluation of the patient with neuromuscular disease must consider not only the neuromuscular deficits, but also the effects the disease may have had on other organ systems, particularly respiratory and cardiovascular. These secondary effects may

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have a significant impact on the administration and course of both general and regional anesthesia in these patients.

Evaluation and documentation of preexisting neurologic deficits is a vital part of the preoperative anesthesia workup for any patient with an underlying neurologic disorder. This is imperative whether regional or general anesthesia is planned. Changes in neurologic status are frequently seen in the perioperative period in these patients, and the documentation of preexisting deficits facilitates the interpretation of any changes seen postoperatively.

The patient with generalized neurologic/neuromuscular disease may be at risk for respiratory compromise in the perioperative period. In particular, impaired ventilatory reserve with reduced ability to respond to hypercapnia and hypoxia may result in an increased risk of respiratory failure.[2,3] The site of surgical incision affects the risk of respiratory complications, with a higher incidence in patients undergoing upper abdominal and thoracic procedures. The method of perioperative analgesia may have a significant influence on this risk of respiratory compromise, providing the anesthesiologist with an opportunity to positively influence the patient's course.

In addition to hypoventilation, dysfunction of the pharyngeal muscles and the potential of aspiration add to the possibility of pneumonia postoperatively. Maintenance of an awake patient can only aid in the prevention of aspiration. Additionally, an endotracheal tube can be protective at the expense of further loss of muscle tone of both the respiratory and pharyngeal muscles. Finally, patients with advanced neurologic disorders can have a degree of restrictive lung disease which makes mechanical ventilation higher risk for barotraumas.[4]

Preoperative assessment of respiratory function and reserve is important and may include measurement of oxygen saturation, arterial blood gas, pulmonary function, and maximum negative inspiratory force. History of swallowing evaluations can be helpful to highlight additional risks in the perioperative period.

Similarly, the cardiovascular effects of neuromuscular disorders must also be considered in the preoperative evaluation. Autonomic dysfunction occurs with many neurologic disorders and constitutes the major contributor to complications related to this organ system. ALS, Guillain-Barré syndrome, multiple sclerosis, and spinal cord lesions above the level of T6 can all have alteration of the autonomic nervous system. In addition, diabetes mellitus, amyloidosis, uremia, and connective tissue disorders are systemic diseases associated with autonomic dysfunction which may coexist in these patients as well.[5,6]

Several findings in the preoperative evaluation may guide the clinician to an increased suspicion for the presence of autonomic dysfunction.[7] The absence of beat-to-beat heart rate variability with deep breathing is one of the most sensitive signs of autonomic dysfunction. Additional characteristic signs include resting tachycardia, orthostatic hypotension, cardiac dysrhythmias, and impotence.

Because of the presence of autonomic dysfunction, these patients have shortening of the Q-T interval which puts them at risk for cardiac conduction abnormalities. In addition, wide fluctuations in blood pressure leads to the frequent need for intraoperative vasopressor therapy. Required avoidance of oral intake makes the presence of relative hypovolemia common. A sympathectomy from neuraxial blockade, but potentially a variable amount from narcotics and inhalational anesthetics as well, can result in exaggerated hypotension in this setting. Finally, unexpected intraoperative cardiorespiratory arrests have been reported in patients with autonomic dysfunction which is second in frequency only to respiratory failure.[8,9]

Myocardial dysfunction and arrhythmias caused by changes in the cardiac muscle and conduction pathways are associated with numerous myopathic diseases including the muscular dystrophies, Guillain-Barré syndrome, and polio. A high index of suspicion must be maintained in the preoperative evaluation of these patients, as exercise tolerance is likely to be very limited by underlying neuromuscular disease. Screening by historical assessment is often inadequate.

Regional Anesthesia and Multiple Sclerosis

Multiple sclerosis is an acquired central nervous system disease characterized by multiple sites of demyelination in the brain and spinal cord. The periventricular white matter, optic nerves, and brainstem are most often affected. Multiple sclerosis does not affect the peripheral nervous system. Demyelination of axons results in a slowing of sensory and motor conduction which leads to widely variable clinical signs and symptoms specific to the sites of demyelination. Examples of the spectrum of symptomatology include the following: visual disturbances because of involvement of the optic nerve, gait disturbances and incoordination secondary to cerebellar changes, and spinal cord demyelination resulting in paresthesias, weakness, and bowel/bladder incontinence.

The diagnosis of multiple sclerosis is made on clinical grounds because there are no specific diagnostic tests. Supportive information may be gained from evoked potential studies with visual, brainstem, and somatosensory potentials revealing slowed conduction. Magnetic resonance and computed tomographic imaging are used to identify plaques throughout the central nervous system. Seventy percent of patients will exhibit a nonspecific increase in protein on cerebrospinal fluid examination.[10]

The clinical course of multiple sclerosis is characterized by a series of remissions and exacerbations over years. Eventually, however, residual symptoms begin to persist between relapses. Extreme variability is seen among individuals, and the waxing and waning course makes it difficult to evaluate the effects of therapeutic interventions. Treatment is mainly supportive with avoidance of stress, excessive fatigue, and marked changes in temperature. Increasing evidence is accumulating that multiple sclerosis is most likely an autoimmune disease involving T cells. Research is directed at treating inflammatory damage to myelin as well as remyelination techniques. Corticosteroids may induce remission during an acute attack[11] but likely do not affect the long-term course of the disease. Interferon β may augment natural disease suppression mechanisms. A decoy similar to the structure of myelin for the autoantibodies has been used. It is called glatiramer, and studies so far are unclear if it is more efficacious alone or in combination with interferon. Intravenous immunoglobulins can reduce the relapse rate, but not likely disease progression. Nonspecific immune suppressants including azathioprine and methotrexate have variable effects. Symptomatic treatment of muscle spasms is also indicated.[12]

There are some case reports that indicate that even intravenous lidocaine has been associated with unmasking the symptoms of multiple sclerosis in a previously undiagnosed patient. Recently, endogenous oligopeptides capable of sodium channel blockade have been discovered in higher concentrations in the cerebrospinal fluid of multiple sclerosis patients compared with normal controls. In fact, these peptides may be active at sites of demyelination and produce some of the negative symptoms of multiple sclerosis, including weakness. Areas of damaged myelin in which ectopic foci of depolarization are capable of producing some of the positive symptoms of multiple sclerosis including spasticity.[13] Furthermore, there is some literature indicating that lidocaine and mexiletine can be used to treat some of the positive symptoms of multiple sclerosis.[14]

The exacerbating factors of stress, fatigue, changes in temperature, and infection are potentially associated with the perioperative period for more than one reason.[15] Delineating the natural course of the disease from the effects of surgery and anesthesia can be very difficult. The purported effects of anesthesia on the course of multiple sclerosis continue to be controversial. There also continues to be a very small number of studies in this area. However, it is the neurologic disease that has the most information about the effects of regional anesthesia. Because of a continuing lack of evidence of safety, reluctance to utilize regional techniques in multiple sclerosis patients persists.

Many of the studies and case reports available involve obstetric patients with multiple sclerosis, which constitutes a subset of patients likely to be considered for regional anesthesia. The natural history of multiple sclerosis in pregnancy is characterized by remission during gestation[16,17] because of a presumed immunomodulatory protective effect.[18] This is also seen in other parturients with autoimmune disorders such as rheumatoid arthritis. In fact, patients who have had a full-term pregnancy have a tendency toward an increased time interval to sustained disability. Patients are likely to have more multiple sclerosis relapses in the first 3 months postpartum regardless of whether they received an epidural.[18]

Neuraxial, and in particular spinal, anesthesia has been implicated as a potential cause of exacerbations in these patients[19] even though contradictory retrospective studies and case reports exist.[20,21] Theories to explain any exacerbation of multiple sclerosis symptoms by spinal anesthesia focus on the potential for an increased susceptibility of demyelinated areas of nerves to the neurotoxic effects of local anesthetics.[20] The three to four times higher concentration of local anesthetic reaching the spinal cord white matter with subarachnoid as opposed to epidural anesthesia could explain the higher risk of exacerbation posed by this modality.[22] Schapira[23] demonstrated that diagnostic lumbar puncture alone did not appear to induce relapses in patients with multiple sclerosis, lending support to the theory that any effects of spinal anesthesia on multiple sclerosis are related to local anesthetic neurotoxicity. In addition, intrathecal morphine has also been used successfully without exacerbation anecdotally in patients with multiple sclerosis.

Bader et al.[20] performed a retrospective and partially prospective review of all obstetric multiple sclerosis patients at the Brigham and Women's Hospital between 1982 and 1987 and noted no significant difference in exacerbation rates between patients receiving epidural anesthesia and local infiltration for vaginal delivery. The total number of pregnancies in patients with multiple sclerosis in this study was 32. However, all of the women who did experience a relapse within 3 months postpartum had received epidural anesthesia with a concentration of bupivacaine greater than 0.25%. This was a total of three patients. The authors proposed that the use of higher bupivacaine concentrations over a longer period of time (i.e., labor epidurals) may affect the rate of postpartum multiple sclerosis relapse, particularly if multiple local anesthetic boluses are required. Warren et al.[24] also reported minor exacerbations in a patient following two separate epidurals (years apart) for vaginal delivery, although a relatively large total dose of bupivacaine was used on the second occasion only. Of note, although these incidents suggest that local anesthetics may potentially produce neurologic symptoms in demyelinated areas of patients with multiple sclerosis, these effects have not been permanent and generally gradual recovery over time is the rule.[25]

Despite these concerns, there are many reports of successful use of epidural anesthesia in multiple sclerosis patients without evidence of relapse. Capdeville and Hoyt[26] performed a retrospective review of all obstetric patients with multiple sclerosis admitted to University Hospitals of Cleveland from 1986 to 1993. Over this 7-year period, eight women with multiple sclerosis underwent eight vaginal deliveries, one cesarean delivery, and five obstetric-related procedures. The anesthetic techniques used were five epidurals, two general anesthetics, one pudendal block, and one narcotic technique. Only two exacerbations of multiple sclerosis were noted by chart review. One of these occurred after a general anesthetic, and the other was noted in a patient receiving a pudendal block. No exacerbations were seen in patients receiving epidural anesthesia.

Crawford et al.[21] documented only one perioperative relapse in 50 nonobstetric and seven obstetric patients with multiple sclerosis receiving lumbar epidural anesthesia. Again, the numbers are too small to lead to generalized recommendations but do indicate anecdotal success without complication involving the use of regional anesthesia in patients with multiple sclerosis.

Confavreux et al.[27] published in 1998 one of the largest prospective studies on the rate of pregnancy-related multiple sclerosis relapses. It included 269 pregnancies in 254 women from 12 European countries from 1993 to 1995. A total of 42 patients received epidural analgesia. The rate of relapse per year was not different when compared with those who did not receive an epidural.

A significant concern in patients with multiple sclerosis is the presence of autonomic dysfunction and the potential for chronic hypovolemia in these immobilized patients. Both factors increase the risk of hemodynamic instability during anesthesia. Kytta and Rosenberg[28] evaluated 56 adult patients with documented multiple sclerosis undergoing a total of 71 anesthetics at the University Central Hospital, Helsinki, Finland, from 1973 to 1982. A retrospective evaluation was made of the different anesthetic techniques used – 42 general anesthetics, five regional anesthetics, and 24 infiltrative techniques. All five patients receiving regional anesthesia (three epidurals and two spinals) had marked hypotension with significant sympathetic blocks. The authors subjectively observed a reduced response to intravenous fluids and vasopressor therapy. They proposed that this may be related to autonomic dysfunction in patients with multiple sclerosis which had been exacerbated by a chronic volume-depleted state.

The use of regional anesthesia in patients with multiple sclerosis remains unclear. Multiple case reports support its successful use, particularly in obstetric patients. Other case reports suggest a risk of perioperative symptom exacerbation and hemodynamic instability. If regional anesthesia is considered, the risk and benefits must be fully discussed with the patient. Special note during these discussions must be made of the potential for exacerbations of multiple sclerosis related to stress and temperature changes associated with the perioperative period regardless of the anesthetic technique used. In addition, parturients have a particular issue with increased incidence of multiple sclerosis relapse early in their postpartum period.

Regional Anesthesia and Amyotrophic Lateral Sclerosis

ALS is a degenerative disease of lower motor neurons, motor nuclei of the brainstem, and descending pathways of upper motor neurons.[29] It results in clinical features of both upper and lower motor neuron lesions with variability depending on the muscle groups involved. ALS limited to brainstem nuclei is referred to as pseudobulbar palsy. Symptoms limited to the motor cortex are then usually referred to as primary lateral sclerosis.[30] The etiology remains unclear, and the disease most frequently affects males in their fifth to seventh decade of life.

The clinical features of ALS involve progressive muscular atrophy with weakness and fasciculations of skeletal muscles. Bulbar muscle weakness often predominates with an associated risk of aspiration. A characteristic emotional lability is seen.[30] Autonomic nervous system dysfunction is common with the associated risk of exaggerated hemodynamic responses during anesthesia. Death from myocardial or respiratory failure ensues, often within 6 years of the onset of symptoms.

Epidural anesthesia has been successfully used in patients with ALS.[31] Kochi et al.[31] reported three cases in which lumbar epidural anesthesia was used, emphasizing the advantage of avoiding tracheal intubations. In this patient population, any duration of mechanical ventilation could accelerate the loss of muscle tone, and weaning from the ventilator could be quite a challenge. However, a high epidural or spinal block can affect intercostals muscle function with detrimental effects in patients with minimal ventilatory reserve.

Regional Anesthesia and Spinal Cord Injuries

Spinal cord injury has classically been divided into two distinct stages. Initial injury is classified as spinal shock which consists of a 1- to 3-week period of flaccid paralysis including loss of sensation temperature regulation, and spinal cord reflexes below the

level of injury.[32] Hypotension, bradycardia, and changes in the electrocardiogram (premature ventricular contractions, nonspecific ST-T wave changes) are characteristic. Regional anesthesia is not frequently used during this stage of spinal shock because of the evolving neurologic injury. It is necessary to be able to follow the examination and the above-mentioned potential of neurotoxicity of local anesthetics in extremely susceptible nerves. There is also a risk of hemodynamic instability as well as hypothermia.

The chronic stage of spinal cord injury is characterized by skeletal muscle spasticity and the return of spinal and autonomic reflexes below the level of injury. Autonomic hyperreflexia is seen in approximately 85% of patients with lesions at or above T6.[33] In this setting, a reflex response may be produced by a cutaneous (incision) or visceral (bladder distension, uterine contraction) stimulus below the level of injury. This afferent stimulus activates preganglionic sympathetic nerves, resulting in severe hypertension because of intense vasoconstriction below the level of the lesion. Under normal conditions, this response is modulated by inhibitory impulses from higher central nervous system centers. With a spinal cord lesion, this inhibitory input is lost and the vasoconstriction proceeds unimpeded. The resulting hypertension stimulates the carotid sinus baroreceptors, leading to reflex bradycardia and vasodilation above the level of injury.[32] As the cord level rises above T6, the potential for compensatory vasodilation decreases and the potential for severe injury can include acute left ventricular failure with pulmonary edema, dysrhythmias, cerebrovascular hemorrhage, retinal bleeding, seizures, and an increasing number of deaths.

Prevention and early treatment of autonomic hyperreflexia is critical. Both general and regional anesthesia have been used effectively. Broecker et al.[34] noted that spinal and epidural anesthesia were logical choices to prevent autonomic hyperreflexia because the afferent limb of the reflex would be blocked. Spinal anesthesia has been shown to be particularly useful,[35] but epidural blocks are less reliable.[34] Parturients at risk for autonomic hyperreflexia from uterine contractions are likely to benefit from the early use of continuous lumbar epidural analgesia after the onset of labor.[36] Baraka[37] reported the successful use of epidural meperidine in a laboring patient at risk for autonomic hyperreflexia. In addition to its prophylactic use, regional anesthesia has been used therapeutically in patients with autonomic hyperreflexia.[36]

Regardless of the anesthetic technique used, medications should be available to treat severe hypertension in the patient with autonomic hyperreflexia. Sodium nitroprusside, ganglionic blockers (i.e., trimethoprim), and alpha blockers (i.e., phentolamine, phenoxybenzamine) have been used.

Concerns often raised regarding the use of spinal anesthetics in this group of patients with spinal cord injury include potential difficulty in placement, difficulty in control or examination of block level, and a potential increased risk of hypotension. Lambert et al.[35] performed a retrospective review of 78 procedures in 50 spinal cord–injured patients considered "at risk" for autonomic hyperreflexia. No significant differences were seen in intraoperative blood pressure between those receiving spinal or general anesthesia. Both techniques seemed to protect equally against intraoperative hypertension.

Regional Anesthesia and Peripheral Neuropathies

Peripheral neuropathies result from either the disruption of axons with distal degeneration or segmental demyelination caused by Schwann cell degeneration.[38] They classically start distally and spread proximally resulting in a "glove and stocking" distribution of decreased sensation, weakness, and reduced reflexes. Some, such as diabetic and alcoholic neuropathy, can be associated with tender muscles. The etiologies of peripheral neuropathies are considerable, including metabolic disorders

(diabetes mellitus, renal failure, hepatic failure, porphyria, and nutritional deficiencies), connective tissue disorders, infection, toxins, malignancy, endocrine disorders, and Guillain-Barré syndrome. Diagnosis depends on metabolic screening tests, serology, infection, and autoimmune evaluations. Electromyogram studies reveal evidence of denervation and a reduction in nerve conduction velocity.

Diabetic peripheral and autonomic neuropathies are encountered frequently in patients presenting for anesthesia and surgery. Clinically, the peripheral neuropathy predominantly affects the lower extremities with paresthesias, weakness, and sensory loss more distally and often worse at night. Occasionally, the neuropathy of diabetes may present as a mononeuropathy causing transient pain and weakness in an isolated nerve distribution. The associated autonomic neuropathy may be significant, with anesthetic implications related to an increased risk of intraoperative hemodynamic instability[39] and an increased risk of unexplained intraoperative cardiac arrest.

The use of regional anesthesia in patients with preexisting peripheral neuropathies depends on a thorough analysis of the risks and benefits for each individual patient. The diabetic patient with a propensity toward perioperative cardiovascular complications[39] might benefit from a regional, particularly spinal, technique that allows the awake patient to report episodes of angina. Another purported advantage of epidural or spinal anesthesia in diabetic patients relates to an improved ability to maintain blood glucose control with the inhibition of the surgical stress response.[40] Certainly, some patients may have exaggerated hypotension with respect to their preexisting autonomic neuropathy. This must be weighed against other risks and benefits that would affect the patient. Furthermore, large doses of local anesthetics have been associated with myocardial depression in diabetic patients.[41]

Guillain-Barré syndrome is an acute inflammatory demyelinating disease of the peripheral nervous system with an incidence of approximately 1:100,000 persons per year.[42] It is postulated to have an autoimmune process directed at myelin similar to multiple sclerosis. Although there may be a correlation with a history of surgery, vaccination, recent respiratory or gastrointestinal tract infection, the etiology is considered to remain unknown. Patients present with the acute onset of lower motor neuron paralysis including flaccid paralysis and reduced reflexes. It begins in the lower extremities and progresses cephalad over hours to days.[43] Bulbar dysfunction and intercostals muscle weakness may ensue, with resultant respiratory failure and the patient's inability to protect their airway. Painful distal extremity paresthesias are common. Autonomic dysfunction occurs in a significant number of patients[44], which results in hemodynamic instability, tachycardia, and cardiac conduction disturbances. Ninety percent of patients will have the most progressive symptoms within 2 weeks.

The treatment of Guillain-Barré is mainly supportive and nonspecific. Many patients will require endotracheal intubation and mechanical ventilation secondary to the respiratory weakness and bulbar dysfunction. Plasmapheresis and intravenous immunoglobulin have been used as treatments during the acute phase and are considered to be equally efficacious. Steroids have been studied and may actually have a deleterious effect. Management of hemodynamic variability associated with autonomic dysfunction can be very challenging. Guillain-Barré usually resolves spontaneously over weeks to months, but approximately 20% of patients will have residual neurologic deficits. The mortality rate is estimated at 5%.[25]

Regional anesthesia has been used successfully in patients with Guillain-Barré syndrome. In particular, epidural anesthesia has been used in parturients with Guillain-Barré without adverse effects.[45] These patients had some residual effects from an episode of Guillain-Barré in the past, but did not have an acute episode of the disease. Epidural narcotics have been used without complication in the acute phase of the disease in an attempt to control painful paresthesias.[46,47] Although the case reports are infrequent, this is another example that narcotics have not been shown to cause toxicity administered neuraxially in patients with neurologic disease[48,49] – even in the setting of acute demyelination. However, no patients received local

anesthetics in the acute phase of Guillain-Barré. When considering regional techniques, patients can have exaggerated responses to indirect vasopressors because of their autonomic dysfunction.

Regional Anesthesia and Muscular Dystrophy

Muscular dystrophies are primary disorders of muscle without clearly evident proximal cause, resulting in progressive weakness. Of the several types, Duchenne's is the most common and most severe.

Duchenne's muscular dystrophy is a sex-linked recessive trait usually only clinically expressed in males. It involves a degeneration of skeletal muscle with atrophy and increased fat and fibrous tissue, but no evidence of denervation. The result is progressive symmetric weakness culminating in death by 15–25 years of age usually attributable to congestive heart failure or pneumonia. Diagnosis is based on muscle biopsy, and increase of serum creatine kinase may be followed to track progression of the disease.

Cardiovascular involvement, although not predominant, may result in significant morbidity and mortality. Congestive heart failure, mitral regurgitation secondary to papillary muscle dysfunction, and sinus tachycardia are common.[50] Dysrhythmias have resulted in cardiac arrest during induction of general anesthesia[51] or in the immediate perioperative period.[52]

Patients with muscular dystrophy are at increased risk for pneumonia and respiratory failure in the perioperative period. Kyphoscoliosis and skeletal muscle weakness produce a restrictive pattern on pulmonary function testing. In addition, depressed laryngeal reflexes and reduced gastric emptying combine to predispose these patients to pulmonary aspiration. Careful monitoring of pulmonary function and attention to clearance of secretions and chest physical therapy are important.

Regional anesthesia may offer significant advantages in these patients. The use of agents known to trigger malignant hyperthermia can be avoided in this population of patients who have an increased risk of this complication.[53] The risk of pulmonary aspiration may be decreased provided that regional anesthesia allows sedation to be minimized. Finally, regional techniques are not without problems. Kyphoscoliosis may lead to technically difficult placement of spinal or epidural blocks. Paralysis of intercostals muscles from regional anesthesia may predispose to respiratory insufficiency.

Regional Anesthesia and Myotonic Dystrophy

Myotonic dystrophy is the most common of the myotonic disorders which include myotonia congenital and the paramyotonias. The disorders are characterized by persistent contraction and delayed relaxation following muscle stimulation which is unrelieved by denervation or paralysis.[54] Myotonic dystrophy is inherited as an autosomal dominant trait with symptoms becoming chronically evident in the second or third decade.[55] There is progressive deterioration of skeletal, cardiac, and smooth muscle function. Initially, involvement of the intrinsic hand and facial muscles progresses to proximal limb musculature as well as bulbar dysfunction with weakness of pharyngeal and laryngeal muscles. Diaphragmatic involvement is common. Cardiomyopathy is common as well. The cardiac conduction system is particularly affected with a significant risk of dysrhythmias and atrioventricular block.[56,57] Bulbar dysfunction and delayed gastric emptying make these patients at high risk for pulmonary aspiration. Associated endocrine disorders also occur, including diabetes mellitus, adrenal, and thyroid dysfunction. Ultimately, death occurs as a result of dysrhythmias, respiratory,

or cardiac failure. Treatment is mostly supportive, but can involve the use of quinine or procainamide for myotonic symptoms.[56]

When patients with myotonic dystrophy present for anesthesia, the preoperative evaluation of pulmonary function is critical. Pulmonary function tests usually reveal a restrictive deficit with mild arterial hypoxemia on a blood gas. A preoperative measurement of baseline negative inspiratory force may be a useful guide to perioperative management. A baseline electrocardiogram should be obtained to assess for cardiac conduction abnormalities. Any underlying respiratory infection should be treated.

General anesthesia presents unique problems in the patient with myotonic dystrophy. Succinylcholine may result in exaggerated contraction of muscles resulting in more difficulty with the placement of an endotracheal tube as well as ventilation. The use of neostigmine to reverse neuromuscular blockade may precipitate myotonic contractions. Patients tend to be extremely sensitive to the respiratory depressant effects of sedatives, opioids, and general anesthetics.[58] An underlying component of central sleep apnea is often present, which further complicates airway management and necessitates added caution in the use of sedatives.

The use of regional anesthesia in patients with myotonic dystrophy is attractive because of the avoidance of neuromuscular blocking agents and their reversal drugs. In addition, the use of sedatives and other anesthetics that may produce respiratory depression can be minimized. Finally, avoidance of triggering agents for malignant hyperthermia can be achieved even though only an association, not a definite risk, has been demonstrated between myotonic dystrophy and this genetic disease.[57] Regional anesthetics can present a different set of concerns. Myotonic contractions are not relieved by spinal or epidural anesthesia – only direct infiltration of an affected muscle with local anesthetic will relieve myotonia. In patients with marginal ventilatory reserve, the effect of high epidural or spinal blockade on intercostals muscle function must be considered, especially because many of these patients may have diaphragmatic dysfunction. When performing regional anesthesia, additional sedatives and anxiolytics should be used with caution. Respiratory status should be continuously assessed for signs of hypoventilation or apnea.

Pregnant patients with myotonic dystrophy may require anesthesia for labor and delivery. General, spinal, and epidural anesthetics have been used successfully in these patients for caesarean delivery.[59,60] However, myotonia and weakness may be exacerbated during pregnancy. Patients with myotonic dystrophy are at increased risk for caesarean delivery because of prolonged labor, as well as postpartum hemorrhage from uterine smooth muscle dysfunction.[59,60]

Cold is a well known trigger for myotonic contractions. Therefore, no matter what technique is used, normothermia is required throughout the perioperative period.[61]

Regional Anesthesia and Myasthenia Gravis

Myasthenia gravis is an autoimmune disorder of unknown etiology affecting the neuromuscular junction with a decrease in the number of acetylcholine receptors and the presence of antireceptor antibodies in 70%–90% of patients.[62] Skeletal muscle weakness is characteristically worsened by activity. Although smooth and cardiac muscle are uninvolved, myocarditis and dysrhythmias may be present.[63] Treatment modalities include cholinesterase inhibitors, corticosteroids, immunosuppressants, plasmapheresis, and thymectomy. Progressive weakness may be associated with progression of the disease (myasthenic crisis) or may reflect excessive muscarinic effects of anticholinesterase drugs (cholinergic crisis). Evaluation of the patient's response to the administration of edrophonium allows differentiation between the two phenomena.

The major anesthetic consideration in myasthenia gravis relates to the use of neuromuscular blockers with affected patients displaying extreme sensitivity to

nondepolarizing blockers is unpredictable. Preexisting skeletal muscle weakness, which is present in varying degrees in these patients, may be potentiated by the relaxant effects of volatile anesthetic agents. Finally, ester local anesthetics may display a prolonged elimination half-life because of reduced plasma cholinesterase activity in patients treated with anticholinesterases,[64] suggesting that amide local anesthetics may be preferable when high or repeated doses are anticipated.

Patients with myasthenia gravis need preoperative assessment of both pulmonary function and aspiration risk because of bulbar dysfunction. Patients with preexisting respiratory compromise are predisposed to significant respiratory depression from anesthetic agents. Therefore, sedatives should be used with caution.

Patients should be monitored closely throughout the perioperative period for myasthenic or cholinergic crisis as well as for gradual deterioration in respiratory function precipitated by stress, infection, missed or excessive anticholinesterase doses, electrolyte abnormalities, or aminoglycoside antibiotics. Identifying patients at particular risk for perioperative compromise and the need for postoperative ventilation was delineated by Leventhal et al.[65] as the following:

1. A history of myasthenia gravis for more than 6 years.
2. A history of unrelated chronic obstructive airway disease.
3. A pyridostigmine dose greater than 750 mg/day during the 48 hours immediately preoperative.
4. A vital capacity of less than 2.9 L.

Regional anesthesia has been used successfully in patients with myasthenia gravis. It is the preferred analgesic technique in laboring parturients with the disease who are planning a vaginal delivery.[66] The use of regional anesthesia may reduce respiratory risk by avoiding the depressant effects of opioids as well as inhaled agents and neuromuscular blockers. In addition, postoperative analgesia and chest physical therapy can also be managed better with neuraxial analgesia. Once again, there is the potential risk of intercostal blockade resulting in respiratory compromise. As is also present in some of the preceding disease states, the combination of bulbar dysfunction with respiratory compromise makes securing the airway with an endotracheal tube somewhat advantageous to avoid potential aspiration.

Conclusion

Regional anesthesia has been used successfully in patients with neurologic disease. The literature is scarce and limited. There are distinct benefits in avoiding the side effects of neuromuscular blockers, general anesthetics, and opioids. The whole patient should be evaluated and examined before any type of anesthetic to document disease progression and effects on other organ systems. An informed decision should be made by the patient and clinician. A regional technique should probably be avoided in the setting of an acute inflammation of the nerves. If a regional technique is used, lower concentrations of local anesthetics should be considered. Lidocaine should be avoided. Neuraxial narcotics with careful attention to dosing and postoperative monitoring may be a safe alternative.

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