The Preoperative Neurological Evaluation

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Abstract

Neurological diseases are prevalent in the general population, and the neurohospitalist has an important role to play in the preoperative planning for patients with and at risk for developing neurological disease. The neurohospitalist can provide patients and their families as well as anesthesiologists, surgeons, hospitalists, and other providers guidance in particular to the patient's neurological disease and those he or she is at risk for. Here we present considerations and guidance for the neurohospitalist providing preoperative consultation for the neurological patient with or at risk of disturbances of consciousness, cerebrovascular and carotid disease, epilepsy, neuromuscular disease, and Parkinson disease.

Keywords

cerebrovascular disorders, neurohospitalist, clinical specialty, epilepsy, movement disorders, neuromuscular diseases

Introduction

Neurological diseases are common, having a life time prevalence of 6% in the general population though varying widely in individual disease prevalence and incidence.^{1,2} Neurohospitalists play an important role in the pre-operative assessment of both patients with and those at risk for the development of neurological disease to help ensure best patient outcomes. The following is a discussion of the preoperative evaluation and considerations for patients with or at risk for common neurological diseases, namely disturbances of consciousness, cerebrovascular and carotid disease, epilepsy, neuromuscular disease, and Parkinson disease (PD).

Disturbances of Consciousness: Delirium and Dementia

Delirium is an acute change in mental status with a fluctuating course of attentional deficits accompanied by either disorganized thinking or changes in the level of arousal.³ Delirium is associated with a 1-year reported mortality rate of 35% to 40%,⁴ an average increase in length of stay of 5 to 10 days,⁵ and a mean increase in the total 1-year health care costs of \$16 000 for hospitalized elderly patients.⁶ Postoperative delirium is common, with an incidence of 10% to 18% following general surgery, 53% following orthopedic surgery, and 74% following open heart surgery,⁷ and the increased mortality risk associated with postoperative delirium extends up to a decade after surgery.⁸ The neurohospitalist plays a vital role in not only assessing patients with prior stroke, other neurologic disease, or prior episodes of delirium but also providing consultation for the prevention of perioperative delirium.

The goal of the preoperative neurological evaluation is to prevent delirium in the first instance. The key aspects of evaluation are the recognition, evaluation, and treatment of a patient's modifiable and nonmodifiable risk factors for delirium; a description of her or his baseline cognitive status; and guidance to mitigate these risks.

The most consistently identified risk factors for delirium are age of ≥ 70 , baseline cognitive impairment, and functional dependence.⁹⁻¹² Increased age entails lower neurologic reserve and increased likelihood of having other medical comorbidities, making the elderly patient more prone to delirium.¹³ Such comorbidities include prior stroke or other neurological diseases, prior episodes of delirium, chronic renal and hepatic diseases, and terminal illness.^{4,12,13} Systemic illness, dehydration, and malnutrition evidenced by abnormal sodium, potassium, and glucose levels, hypermagnesemia, blood urea nitrogen (BUN)/creatinine ratio of ≥ 18 , and hypoalbuminemia are risk factors for delirium.^{4,9,10,13} Sensory impairment, namely vision and hearing, puts a patient at risk for delirium.^{10,11} Active alcohol abuse and depression, preoperative psychotropic drug use, and the presence of psychopathological symptoms are of small to moderate

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Table 1. Classes of Medications Associated With Delirium.^{7,19}

Significant Association	Less Clear Association
Opioids	Histamine (H2) blockers
Antihistamines	Tricyclic antidepressants
Benzodiazepines	Nonsteroidal anti-inflammatories
Anticholinergics	Dihydropyridine calcium-channel blockers
	Antiparkinsonian

correlation with delirium incidence.^{9,12} Several delirium prediction tools have been developed for cardiac noncardiac and orthopedic surgeries, which may prove useful to the consulting neurohospitalist.^{9,14,15}

Cognitive impairment and dementia warrant particular consideration, given their strong association with postoperative delirium.^{9,10,12,13} Unlike delirium, dementia is a slowly progressive chronic process with little fluctuation and no change in the level of consciousness until late in its course.¹⁶ A baseline cognitive status assessment that can be readily repeated by nonneurologist providers is invaluable for delirium monitoring throughout the perioperative period. Various bedside assessments have been used as gross assessments, including clock-drawing tasks, the Telephone Interview for Cognitive Status (TICS), the Mini-Mental Status Examination (MMSE), and recounting the days of the week in reverse.¹⁶ For instance, a baseline MMSE of <24/30 has been associated with an increased risk for postoperative delirium.¹¹ An understanding of the patient's baseline independence in activities of daily living would also be of value postoperatively as preparations are made for discharge home or for rehabilitation.

If a patient is noted to have cognitive impairment or dementia, a thorough investigation for reversible etiologies should be undertaken in the outpatient setting prior to surgery. This would include consideration for and evaluation of hypothyroidism, thiamine deficiency, vitamin B12 deficiency, infections, medication toxicity, drug and alcohol use, subdural hematomas, normal pressure hydrocephalus, and intracranial tumors.¹⁷ If no reversible cause of cognitive impairment is identified, patients and their families should be counseled that postoperative delirium is likely, with cognitive recovery likely to be prolonged or incomplete following major surgery.¹⁸

Medications are a key consideration in preoperative planning, given their associated risk for withdrawal syndromes and delirium, particularly in the elderly and cognitively impaired individuals (Table 1).^{7,19} Antiparkinsonian medications such as monoamine oxidase inhibitors, amantadine, catechol-O-methyltransferase inhibitors have been implicated.^{7,19} Digoxin and antipsychotic use, however, do not clearly contribute to an increased risk.¹⁹ Patients are at risk for summed anticholinergic effects in the setting of outpatient polypharmacy (Table 2).^{10,21} Selective serotonin reuptake inhibitors should be continued to avoid delirium due to serotonin

Table 2. Commonly Prescribed Medications With Anticholinergic Activity.^{20,21}

Beta-blockers
Propranolol
Metoprolol
Timolol
Calcium-channel blockers
Nifedipine
Diltiazem
Diuretics
Furosemide
Hydrochlorothiazide
Histamine (H2) blockers
Cimetidine
Ranitidine
Other
Captopril
Codeine
Digoxin
Ibuprofen
Insulin
Isosorbide dinitrate
Nitroglycerin
Prednisolone
Salicylic acid
Warfarin

reuptake inhibitor discontinuation syndrome.²² Neuroleptics are commonly used in the chronic treatment of patients with agitated alterations in behavior, and these should be documented with advisement of the risk for acute dystonic reactions if abruptly discontinued or if continued, the risk for neuroleptic malignant syndrome with general anesthesia.^{18,21} Patients chronically treated with benzodiazepines are at risk for withdrawal and associated seizures if these are abruptly ceased in the perioperative period.²¹ The initiation of regularly scheduled psychoactive medications while hospitalized should be discouraged.⁴

Finally, nonpharmacologic interventions have demonstrated efficacy in preventing delirium in the elderly individuals while hospitalized (Table 3),^{11,23} and the neurologist evaluating a patient preoperatively should comment on these when appropriate. In a group of elderly patients treated with these and other interventions when appropriate, only 10% developed delirium compared to 15% in a nonintervention control group¹¹; while in a community hospital setting similar interventions led to a 35% relative reduction in delirium.²³

Delirium is common and possibly preventable in an estimated 30% to 40% of cases and is currently used as a measure of care quality and patient safety by the National Quality Measures Clearinghouse, Agency for Healthcare Research and Quality (<http://www.qualitymeasures.ahrq.gov/>).⁴ In addition, Medicare also already uses rate of delirium as a quality measure for nursing home residents (<http://www.medicare.gov/> accessed October 8, 2012). The neurohospitalist is well

Table 3. Nonpharmacologic Interventions Effective in Preventing Delirium in the Hospitalized Elderly Individuals.^a

Targeted Risk Factor	Intervention
Cognitive impairment/disorientation	Orientation protocol: Board in room with team member's names and schedule for day Communication to reorient to surroundings Therapeutic Activities: Cognitively stimulating activities 3 times daily
Sleep deprivation	Nonpharmacologic protocol: Warm drink (milk or herbal tea) at bedtime Relaxation tapes or music Back massage Sleep-enhancement protocol: Unit-wide noise reduction strategies Rescheduling medications and procedures to promote sleep
Immobility	Early-mobilization protocol: Ambulation or active range-of-motion exercises 3 times daily Minimal use of immobilizing equipment (eg, bladder catheters, physical restraints)
Visual impairment	Vision protocol: Visual aids (eg, glasses or magnifying lenses) Adaptive equipment (eg, large illuminate telephone key pads, large-print books, fluorescent tape on call bell) With daily reinforcement with adaptations
Hearing impairment	Hearing protocol: Portable amplifying device Earwax disimpaction Special communication techniques With daily reinforcement of adaptations
Dehydration	Dehydration protocol: Early recognition of dehydration and volume repletion (ie, encourage oral intake of fluids)

^a Modified from Inouye and colleagues.¹¹

positioned to not only serve as a consultant but also lead in hospital-wide changes to address delirium rates, such as the early identification of at patients at risk for delirium and protocols for its prevention.

Cerebrovascular and Carotid Disease

Although rare, stroke is a feared surgical complication, with a mortality rate of up to 26%.²⁴ The majority of perioperative strokes are embolic.²⁵⁻²⁸ Hypoperfusion is an uncommon mechanism of perioperative stroke, even in patients with carotid artery stenosis.^{29,30} Less common mechanisms of perioperative stroke include hemorrhagic infarction; air, fat, and paradoxical embolism; and arterial dissection due to neck manipulation.^{25,31}

Table 4. Risk of Perioperative Stroke After Various Surgical Procedures.^{25,32,33,40-42}

Procedure	Risk of Stroke (%)
General surgery	0.08-0.7
Peripheral vascular surgery	0.8-3.0
Isolated CABG	1.4-3.8
Isolated valve surgery	4.8-8.8
Resection of head and neck tumors	4.8
Carotid endarterectomy	5.5-6.1
Combined CABG and valve surgery	7.4
Aortic repair	8.7
Double- or triple-valve surgery	9.7

Abbreviation: CABG, coronary artery bypass grafting.

The risk of perioperative stroke depends on patient characteristics^{26,29,32}; the nature and timing of the procedure^{25,26,32-35}; and certain complications in the postoperative period, including postoperative atrial fibrillation^{36,37} and hypercoagulability.^{35,38,39} This section will focus on the assessment and modification of patient-related (ie, preoperative) risk factors. By and large, preoperative risk factors are a reflection of the traditional risk factors for stroke in the general population. Female sex, renal insufficiency, pulmonary disease, and preoperative discontinuation of antithrombotic therapy are additional preoperative risk factors.³¹

The risk of stroke following general surgery is low (Table 4).³² By contrast, cardiac, vascular, and neurologic procedures are associated with higher risk,^{24-26,40-42} reaching 8% for combined procedures (eg, combined coronary artery bypass grafting [CABG] and valve surgeries)^{25,42,43} and 13% for patients with prior stroke or transient ischemic attack (TIA).²⁹

No validated models are currently available for the prediction of perioperative stroke risk in general surgical patients. A predictive model for assessing the risk of stroke in patients undergoing CABG does exist, using 7 preoperative variables of varying weights to arrive at an aggregate estimate of risk (Table 5).⁴⁴ This risk must be considered in the context of additional intraoperative and postoperative risk factors and weighed against the benefits of surgery.

Many preoperative risk factors, such as advanced age and history of cerebrovascular disease, are nonmodifiable. Nonetheless, a detailed history can meaningfully alter preoperative management. A history of cerebrovascular disease is a strong predictor of perioperative stroke.^{26,29,32-34,42,45} As cerebrovascular reserve remains tenuous in the days and weeks following a stroke, at least 6 months is needed following a stroke or TIA in order to ensure stabilization of the patient's hemodynamic and neurologic status before an elective procedure. Specific questions must be asked to uncover any previously unreported, unrecognized, or inadequately evaluated cerebrovascular symptoms within this 6-month period. Medical management of other preoperative risk factors, such as diabetes and hypertension, must also be optimized.

Table 5. Prediction of Perioperative Stroke Risk in Patients Undergoing CABG.^a

Risk Factor	Weighted Score
Advanced Age	
55-59 years	1.5
60-64 years	2.5
65-69 years	3.5
70-74 years	4.0
75-79 years	4.5
≥80 years	5.5
Nonelective surgery	
Urgent	1.5
Emergent	2.5
Female sex	1.0
Ejection fraction <40%	1.5
Vascular disease ^b	2.0
Diabetes mellitus	1.5
Creatinine >2mg/dL or dialysis	2.0
Total Score	Stroke Risk (%)
0-1	0.3
2	0.4
3	0.6
4	0.8
5	1.0
6	1.2
7	1.6
8	2.1
9	2.9
10	5.8
11	9.8
12	>10.0

Abbreviations: CABG, coronary artery bypass grafting; TIA, transient ischemic attack.

^a Adapted from Charlesworth and colleagues.⁴⁴

^b Includes history of stroke, TIA, carotid stenosis or bruit, or vascular surgery, including leg amputation.

Potentially modifiable preoperative risk factors include carotid artery stenosis, atheromatous aortic disease, and perioperative discontinuation of antithrombotic therapy. Currently there is no evidence to support prophylactic carotid revascularization in general surgical patients.⁴⁶ However, patients with symptomatic carotid artery stenosis (ie, attributable stroke or TIA in the preceding 6 months) have a high risk of perioperative stroke^{29,47} and may benefit from revascularization before or during major cardiac or vascular surgery.⁴⁸ If there is an urgent indication for cardiac surgery, a combined or reversed-stage approach may be used, although the combined approach is likely associated with higher morbidity.⁴⁹

For asymptomatic carotid artery stenosis, the approach should be individualized. The presence of an asymptomatic bruit is not correlated with the severity of the underlying stenosis, nor does it predict perioperative stroke risk.⁵⁰ Therefore, routine revascularization is not warranted in these patients.^{29,47,48} Generally, unilateral asymptomatic carotid artery stenosis of <50%

carries a 2% overall perioperative stroke risk in patients undergoing CABG. The risk increases to 3% for unilateral asymptomatic carotid artery stenosis of 50% to 99%, 5% for bilateral stenosis of 50% to 99%, and 7% for carotid artery occlusion.²⁹ However, 60% of perioperative strokes in these patients are attributable to unrelated mechanisms, suggesting that asymptomatic carotid artery stenosis may be a vascular disease marker rather than a perioperative stroke cause. Nonetheless, patients with high-grade and hemodynamically significant asymptomatic carotid artery stenosis may benefit from revascularization, especially if bilateral. The preoperative evaluation should include brain imaging to rule out silent ipsilateral infarcts, which may strengthen the argument for revascularization prior to surgery in such patients. Transcranial Doppler ultrasonography and computed tomographic or magnetic resonance angiography (CTA and MRA, respectively) may be used to assess silent intracranial microemboli, intracranial blood flow, and the hemodynamic significance of the stenosis,^{51,52} but the utility and validity of these techniques remain to be established.

The identification of a focal atheromatous aortic lesion by transesophageal echocardiography or intraoperative epiaortic ultrasound may support the use of intra-aortic filtration and guide aortic cannulation or clamping during cardiac surgery. The identification of a diffuse aortic plaque burden may influence the decision to pursue an “off pump” approach during cardiopulmonary bypass.⁵³

The perioperative discontinuation of antithrombotic therapy increases stroke risk, particularly in patients with coronary artery disease, but this must be weighed against the risk of surgical bleeding.^{54,55} Current evidence supports the discontinuation of antiplatelet agents at least 5 days prior to CABG and reinitiation of antiplatelet therapy as early as 6 hours postoperatively.⁵⁶⁻⁶¹ By contrast, the safety and effectiveness of continuing antiplatelet therapy in the perioperative period for patients undergoing carotid endarterectomy are better established.⁶²⁻⁶⁶

It is generally considered safe to continue anticoagulation in most patients undergoing dental procedures, cataract surgery, arthrocentesis, and diagnostic endoscopy with or without biopsy.⁶⁷ Continuing moderate-dose warfarin therapy (international normalized ratio [INR] 1.8-2.1) is safe and effective in patients undergoing knee or hip replacement surgery.⁶⁸ For more invasive procedures, the rate of perioperative thromboembolic events in high-risk patients on long-term anticoagulant therapy is 0% with bridge therapy using intravenous heparin and 0.6% in the absence of bridge therapy.⁶⁷ Thus, current evidence supports the continuation of anticoagulation for some invasive procedures, such as colonoscopy and minor surgeries. If oral anticoagulation must be stopped, bridge therapy with heparin should be used and anticoagulation should be resumed early in the postoperative period.⁶⁹⁻⁷¹

Epilepsy

Epilepsy affects approximately 1% of the population.⁷² A significant number of patients undergoing surgery will have

Table 6. Interactions of Common Antiepileptic Drugs With Common Perioperative Medications.^{65,76}

Cytochrome P450 Activity	Effect on Serum Concentrations of Medications	Commonly Affected Medication Classes
Inductors	Decrease	Certain antibiotics Immunosuppressants Beta-blockers Calcium-channel blockers
Phenytoin		
Phenobarbital		
Carbamazepine		
Oxcarbazepine		
Eslicarbazepine		
Inhibitor	Increase	
Valproate		

a history of seizures, prompting neurological consultation regarding risk of perioperative seizures, management of antiepileptic drugs (AEDs), considerations for anesthesia and surgery, and potential postoperative complications.

The risk of perioperative seizures is proportional to the frequency of seizures at baseline.⁷³ Ideally, seizure control is optimized as an outpatient before surgery; however, epilepsy is often intractable and there is always a potential for seizures. Multiple surgical factors increase the risk for seizures, such as alterations in maintenance AED therapy via absorption or scheduling changes, physical and psychological stressors as well as the use of potentially pro-convulsive anesthetics.

It is preferable to maintain AED therapy as close to baseline as possible perioperatively. Despite common instructions not to take anything by mouth the morning of surgery, with the anesthesiologist's and surgeon's approval, patients with epilepsy could be instructed to take their regularly scheduled morning AEDs with sips of water. Baseline oral AEDs should be reinitiated as soon as feasible after surgery. If enteral options are not available, there are intravenous (IV) equivalences for some AEDs (eg, phenytoin, valproate, phenobarbital, levetiracetam, and lacosamide). Perioperative seizures often occur when serum AED levels are low.^{73,74} Routine serum levels probably do not need to be drawn unless prolonged AED withdrawal or significant changes are expected. For example, valproate concentrations can be decreased by certain beta-lactam antibiotics.⁷⁵

There are more than a dozen commonly used AEDs, with varying mechanisms of actions. Many interact with other medications that may be administered during the perioperative period (Table 6).^{65,76} Patients on warfarin should have their INR carefully monitored perioperatively. Valproate is known to affect multiple hematological processes that may lead to increased risk of bleeding (Table 7),^{77,78} though no perioperative adjustment of valproate is needed.^{79,81}

Epilepsy can be treated with modalities other than medications. Patients with epilepsy may have vagal nerve stimulators (VNSs) which can be temporarily turned off, but their presence might limit imaging options such as MR imagings. Other patients may be on the ketogenic diet. They may safely

Table 7. Hematologic Processes Affected by Valproate.¹²⁹⁻¹³²

Thrombocytopenia
Inhibition of platelet aggregation
Reduction of von Willebrand factor
Fibrinogen depletion

undergo general anesthesia but require carbohydrate-free solutions and monitoring of serum electrolytes, glucose, and particularly pH, as they are at risk of metabolic acidosis.^{82,83}

While treatment of seizures may have an impact on surgery, surgery has an impact on seizures. Certain medications used by anesthesiology may influence the threshold for seizures.^{84,85} Generally, lower dosages of anesthetics (eg, etomidate) tend to be pro-convulsive, while higher dosages tend to be anticonvulsive.^{86,87} Exceptions are opioids that tend to be only pro-convulsive. However, the majority of seizures that occur perioperatively are not related to either local or general anesthesia but to the underlying epilepsy.^{73,88,89} The frequency of perioperative seizures in these studies is between 2% and 6%. Also, patients with epilepsy may have psychogenic nonepileptic convulsions, which may increase perioperatively.⁹⁰ Additionally, recovery from anesthesia may lead to transient shivers and myoclonus.

Patients with epilepsy do have more complications than those without a history of seizures. They tend to have more preoperative comorbidities and are at increased risk of postoperative complications, including pneumonia, septicemia, acute renal failure, bleeding, wound infections, and most significantly stroke with no difference in mortality rates.⁹¹

Neuromuscular Disease

Neuromuscular patients can experience various, potentially life-threatening perioperative complications. The primary goal of preoperative evaluation is to predict and minimize potential neurologic complications and enhance the surgical outcomes of patients with neuromuscular disorders.

Respiratory weakness is a common feature of neuromuscular disease, and a careful history to recognize symptoms of reduced function capacity can be invaluable. A history of orthopnea, mild to moderate dyspnea exacerbated by flexing at the back, insomnia, chronic fatigue, or narcolepsy should be investigated. Physical examination should include auscultation of lungs, inspection of accessory respiratory muscle use, and diaphragmatic excursion as well as documented functional vital capacity (FVC) with spirometry. Although there is no clear guideline, pulmonary function test (PFT) and arterial blood gas analysis (ABGA) should be considered as they have been shown to predict pulmonary complications after surgery.⁹² Patients with chronic respiratory weakness are at risk of developing CO₂ narcosis if oxygenation without adequate ventilation is administered during the postoperative period; therefore, aggressive use of noninvasive positive

pressure ventilation (NPPV) with mechanical insufflator-exsufflator, such as CoughAssist,⁹³ is recommended after surgery.

Dysphagia is another common symptom of neuromuscular disease, with a prevalence of up to 62% after endotracheal intubation.⁹³ As neuromuscular patients can have bulbar weakness, a swallowing study for silent aspiration should be considered prior to general anesthesia. If the patient already has a percutaneous endoscopic gastrostomy (PEG) tube, PEG tube feeding should be stopped 6 to 8 hours prior to general anesthesia.

In certain conditions, such as hereditary neuropathy with liability to pressure palsy (HNPP), careful surgical position is a key consideration to avoid entrapment neuropathies. Common entrapment sites include the fibular head and cubital tunnel. When evaluating patients with autonomic nerve dysfunction, it is important to inform the primary team about possible changes in vital signs depending on positioning during surgery.

Certain drugs are known to worsen symptoms of some neuromuscular disorders. Aminoglycoside antibiotics impair neuromuscular transmission and can cause significant weakness.⁹⁴ High-dose steroids are associated with an increased risk of critical illness myopathy after surgery, particularly when nondepolarizing neuromuscular junction blocking agents are used for intubation.⁹⁵ Early mobilization and aggressive postoperative treatment reduce the risk of critical illness myopathy.^{96,97}

Focusing on particular neuromuscular diseases, amyotrophic lateral sclerosis (ALS) and other motor neuron diseases are commonly characterized by bulbar and respiratory weakness, increasing the risk of aspiration pneumonia and various pulmonary complications. Nondepolarizing neuromuscular blocking agents cause prolonged and pronounced neuromuscular blockade and therefore should be used with caution.⁹⁸ General anesthesia combined with epidural anesthesia has been used without complications.

Patients with Charcot-Marie-Tooth disease (CMTD) tolerate anesthesia with few complications.⁹⁹ As denervation predisposes to potassium release, patients with CMTD are theoretically at risk of hyperkalemia with the use of succinylcholine, and it has been used without adverse effects.⁹⁹ Nondepolarizing neuromuscular blocking agents have been used safely, although there are case reports of prolonged weakness related to certain agents.¹⁰⁰⁻¹⁰²

Patients with muscular dystrophies not only have a high prevalence of cardiac involvement but are also at risk of developing perioperative respiratory complications. Preoperative cardiac assessment with electrocardiogram and echocardiography is necessary. Continuous cardiac Holter monitoring should also be considered if arrhythmia is suspected. In addition, careful preoperative pulmonary assessment, including pulmonary function studies, is crucial. Preoperative pulmonary function predicts postoperative respiratory complications in muscular dystrophy patients who are undergoing scoliosis surgery and

have forced vital capacity (FVC) <40%.¹⁰³ Postoperative continuous positive airway pressure (CPAP) and bilevel positive airway pressure (BiPAP) have demonstrated efficacy in these cases.

Although there has been concern of malignant hyperthermia in patients with dystrophin-related muscular dystrophies, there is no evidence to suggest increased risk of malignant hyperthermia during surgery.¹⁰⁴ Succinylcholine is contraindicated in these patients because of the potential risk of rhabdomyolysis and life-threatening hyperkalemia due to sarcolemmal membrane instability.¹⁰⁴ Patients with Duchenne muscular dystrophy are particularly sensitive to nondepolarizing neuromuscular blocking agents.¹⁰⁵

Because of various comorbidities such as diabetes, periodic myotonia, progressive myopathy, and cardiac anomalies, patients with myotonic dystrophy can potentially develop a variety of complications related to surgery. Their frequency of complications is 8.2%, most of which are pulmonary.¹⁰⁶ There is a high incidence of arrhythmia in patients with myotonic dystrophy, necessitating careful perioperative cardiac monitoring. Cold temperature can trigger and/or aggravate myotonia, which can complicate various anesthetic procedures including endotracheal intubation.¹⁰⁷ The myotonic reaction can be treated with phenytoin (4-6 mg/kg/d) or quinine (0.3-1.5 g/d).¹⁰⁸ Patients with myotonic dystrophy are sensitive to certain anesthetics and muscle relaxants, such as succinylcholine, methohexital, etomidate, propofol, and neostigmine.¹⁰⁸ Finally, patients can develop intestinal pseudo-obstruction due to smooth muscle atrophy.

Although many patients with myasthenia gravis achieve disease remission with the use of immunosuppressants, they can experience potentially life-threatening complications in the perioperative period. Patients and their families should be informed about the possibility of myasthenia crisis and/or prolonged ventilator use. In general, patients should keep taking anticholinergic medications as well as immunosuppressants. Most immunosuppressant drugs do not interact with anesthetic medications, except for azathioprine, which may prolong the effect of succinylcholine and inhibit nondepolarizing neuromuscular blockers. If possible, severe myasthenia gravis should be stabilized prior to surgery using short- to intermediate-term treatment strategies, such as intravenous immunoglobulin or plasmapheresis. Pyridostigmine and other anticholinergic medications can theoretically interfere with neuromuscular blocking agents, delaying their effect onset. Considering the risk of respiratory complications, the value of discontinuing pyridostigmine is questionable.¹⁰⁹ Volatile anesthetic agents can be safely used in patients with myasthenia gravis, reducing the need for neuromuscular blocking agents.¹⁰⁸ In postoperative period, reconstitution of pyridostigmine—either oral or its parenteral equivalent form—can be done with half of the preoperative dose given 12 hours after the procedure, increasing to the full dose over the next 2 days.¹¹⁰ For patients who have been on steroids, a boosting dose of IV methylprednisone can also be considered prior to

Table 8. Common Perioperative Medications Which Worsen Myasthenia Gravis.¹¹¹

Antibiotics
Aminoglycosides
Fluoroquinolones
Macrolides
Penicillin
Tetracyclines
Sulfonamides
Clindamycin
Nondepolarizing neuromuscular blocking agents
Pancuronium
Vecuronium
Atracurium
Beta-blockers
Propranolol
Atenolol
Metoprolol
Antiarrhythmics
Procainamide
Quinidine
Local anesthetics
Procaine
Xylocaine
Other
Magnesium

extubation. Finally, surgical and medical team members should be reminded that certain medications could worsen and/or trigger myasthenia crisis (Table 8).¹¹¹

A final neuromuscular disorder with perioperative considerations is malignant hyperthermia syndrome (MHS). This syndrome is characterized by severe muscle rigidity, fever, and cardiac arrhythmia. MHS is now known to be autosomal dominant inherited disorder associated with ryanodine calcium channel genes and thought to be caused by unregulated calcium release from sarcoplasmic reticulum. MHS occurs more with certain anesthetics, such as halothane and depolarizing muscle relaxants. MHS has been reported in various muscle diseases, such as channelopathies, central core disease and mini-core disease (Table 9).^{112,113} The diagnosis is based on clinical presentation and should be suspected when patients develop masseter contractures and muscle rigidity with fever, acidosis, and tachycardia. Genetic analysis has limited value since patients with unknown mutations can still develop MHS.¹¹⁴ Preoperative dantrolene should be considered in at risk patients.

Parkinson Disease

Parkinson disease is a prevalent¹¹⁵ and highly disabling neurodegenerative disease with profound public health impact. While there is no proven cure or prevention for PD, patients are treated with multiple medications to manage PD's motor and nonmotor manifestations and complications. Many patients with later-stage PD have had deep brain stimulation

Table 9. Disorders Associated With Malignant Hyperthermia Syndrome (MHS).^{112,113}

Family history of MHS
King-Denborough syndrome
Evans myopathy
Central core disease
Mini-core disease
Periodic paralysis
Myotonia congenital
Carnitine palmitoyltransferase (CPT) deficiency
Mitochondrial myopathies

(DBS) surgery, and the implanted DBS hardware is a major source of concern to health care providers unfamiliar with this treatment modality. The proper use of these medications and complications of therapy represent a particular challenge for patients with PD undergoing surgery.

Obtaining preoperative images of the best quality poses a particular concern for patients with PD as dyskinetic movements may limit image resolution, and the presence of DBS hardware is a concern for various modalities. For patients without DBS, there are no PD-specific restrictions on imaging procedures. Patients with PD may rapidly fluctuate between the “on” state in which they can easily move, the “on” state with choreiform dyskinetic movements, and the “off” state in which they are bradykinetic and rigid.¹¹⁶ It is recommended to perform imaging studies during the “on” time without dyskinesias. For patients on a regimen requiring frequent medication administration, timing of imaging procedures should help minimize nonadherence to scheduled dosing times.

For patients with DBS, diagnostic ultrasounds, CT scans, and X-rays are generally safe.¹¹⁷ Brain MRI in patients with DBS is of low risk when specific guidelines are followed.^{118,119} The safety concerns associated with MRI include brain tissue heating and subsequent edema at the electrode site as well as device malfunction in the strong magnetic field. While there have been isolated reports of severe adverse events following brain MRI in individuals with DBS,¹²⁰ the guidelines from the US manufacturer of DBS hardware (Table 10) were not followed.¹²⁰ There is less evidence regarding the safety of MRIs of other parts of the body in individuals with DBS, though limited body MRIs have been performed safely in at least 170 patients.¹¹⁸ Medtronic states that MRI is contraindicated if it uses a full-body transmit radiofrequency coil, a receive-only head coil, or a head transmit coil that extends over the chest area.¹¹⁷ There are informal reports of a tentative plan to relax the absolute safety ban on body MRIs in the future.

The primary difficulty in managing patients with PD in the operating room is the timing of their PD medications. Abrupt cessation of levodopa has been associated with parkinsonism–hyperpyrexia syndrome, which involves hyperpyrexia, dysautonomia, and increased creatine kinase.¹²¹ To avoid this syndrome as well as the much more common debilitating “off”

Table 10. Deep Brain Stimulator Manufacturer Recommendations for MRI.¹¹⁷

1. Use only a 1.5 T MRI system
2. Use only a transmit/receive RF head coil
3. Limit displayed average head SAR to no more than 0.1 W/kg
4. Limit the gradient switching (dB/dt) to less than 20 T/s
5. Instruct patients prior to MRI to notify the MRI technician immediately of any unusual sensations
6. Assess patient's status throughout the MRI
7. Stop MRI immediately if the patient becomes unresponsive to questions or experiences any heating, pain, shocking sensations, uncomfortable stimulation, or unusual sensations

Abbreviations: MRI, magnetic resonance imaging; RF, radiofrequency; SAR, specific absorption rate; DBS, deep brain stimulation.

^a Use of these guidelines assumes that the DBS electrodes are connected to subclavicular Implantable pulse generator (IPG), the volts (or amperes) have been turned to 0 and the DBS is turned off. There are additional precautions that must be used if the individual does not yet have the IPG implanted.

times, patients should be allowed to continue their levodopa medication up to the moment of the surgery. Levodopa should then be restarted as soon as possible after the surgery.¹²²

The realities of surgery, including necessary no oral intake requirements, often make these recommendations difficult to follow. Practitioners in Europe demonstrated the efficacy of temporarily replacing levodopa with rotigotine, a transdermal patch, during the perioperative period.¹²³ This medication was discontinued in the United States in 2008 but was reintroduced into US markets in July 2012. Alternatives include nasogastric tube placement or use of an orally disintegrated formulation carbidopa/levodopa. In addition, while not yet approved in the United States, levodopa/carbidopa intestinal gel is a current option for many practitioners. In this treatment, the medication is administered directly to the small intestine through a surgically placed tube. In theory, this delivery method would allow for continuous infusion even during some surgeries. However, we could not identify any research regarding the safety or efficacy of this continuous delivery during surgery. Intravenous formulations of levodopa remain experimental and are limited to only a few centers.¹²⁴ Possibly helpful perioperatively when it is not possible to follow a levodopa regimen with frequent dosing as usually needed for patients with advanced PD, a longer acting formulation's new drug application (http://www.impaxlabs.com/our_divisions/impax_pharma_branded) is currently under FDA review for approval in the United States.

There are three additional difficulties in managing patients with PD in the perioperative period. First, patients with PD may be taking monoamine oxidase-B inhibitors (MAO-Bs, selegiline and rasagiline) that may lead to a fatal serotonergic syndrome when combined with meperidine or dextromethorphan. Some argue that if meperidine and dextromethorphan are not used, surgery is safe in individuals taking MAO-Bs.¹²⁵ However, there are additional interactions between general MAO-B medications and other anesthetics, leading

to the recommendation to discontinue all MAO-Bs at least 3 weeks prior to surgery.¹²⁶ Generally, MAO-Bs do not provide a large symptomatic boost to most patients with PD, and there is no concern of rebound or withdrawal phenomena even when abruptly discontinued. However, we would recommend discussing with the patient's neurologist the discontinuation of the medication prior to elective surgery. Second, both PD and levodopa treatment are associated with hypotension. With proper monitoring and balancing of antihypertension medications, such complications can usually be mitigated.¹²⁶ Finally, surgery can proceed safely even for individuals with DBS. For minor procedures, the stimulation should be set to 0 and the implanted pulse generator (IPG) turned off, though this should be discussed with the prescribing or implanting clinicians.¹¹⁷ This will limit the risk to the IPG during the procedure. Immediately after completion of the procedure, the DBS may be turned back on. For DBS-related perioperative questions, the US manufacturer/manufacturers of DBS hardware may also be contacted.

Individuals with PD have longer hospital stays and increased risk of urinary tract infections, aspiration pneumonia, and bacterial infections in the postoperative period compared to individuals without PD.¹²⁷ Management of patients with PD in the postoperative period works to mitigate these differences to include delirium management, medication timing, medication administration, and appropriate use of anti-nausea medication. For patients who are delirious and PD medications are suspected to contribute, ancillary PD medications may be discontinued (eg, amantadine, MAO-B inhibitors, COMT inhibitors, and anticholinergics) while dopaminergic medications should be continued, especially levodopa, if possible.

In addition to addressing PD patients' delirium in the postoperative period following guidelines discussed earlier in this article, it is important to strictly manage the patient's medication schedule. Conventional guidelines allowing for medications within a specific time period on the wards are not acceptable in patients with PD.¹²² Practitioners are encouraged to work with nursing staff to ensure on-time medication administration. For more details and tips about maintaining strict dosing schedules for patients with PD, readers are encouraged to visit the National Parkinson Foundation Web site about this issue at <http://www.parkinson.org/Improving-Care/Education/Education-For-Patients/Aware-in-Care>.

Medication administration is complicated by postoperative swallowing concerns as well as the need for complete bowel rest in some surgeries. Rotigotine transdermal patch, nasogastric tube administration of levodopa, and orally disintegrating levodopa are options in the postoperative period. Some centers may have IRB-approved availability of IV levodopa, which is an experimental option for the perioperative period as well as critically ill and otherwise incapacitated patients. Injectible apomorphine is not advised as a standing substitution for levodopa or other dopamine agonists. Consultation with the local movement disorder specialists to review this

and the other parenteral options (sublingual, transdermal) is encouraged.

Finally, when addressing the postoperative nausea of patients with PD, practitioners should use either ondansetron or trimethobenzamide. The other antiemetic medications can have antidopamine effects, either through dopamine receptor antagonism or through other mechanisms. This further reduction of the PD patient's already low dopamine supplies exacerbates their PD symptoms.¹²⁸

Conclusion

Neurological diseases are common, with a variety of considerations for perioperative and surgical planning. The key for the neurological evaluation prior to surgery is to provide guidance for the neurohospitalist to help avoid neurological complications, especially those anticipated for a neurological patient's particular diagnosis, and to prevent new neurological disorders from occurring in the perioperative period.

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