

Anaesthetic considerations for patients with pre-existing neurological deficit: *are regional techniques safe ?*

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If in patients with pre-existing neurological conditions any changes or deteriorations are noticed after surgery, anaesthetic drugs or techniques selected are not infrequently blamed. However, the cause of postoperative deficit is not easy to determine and may occur due to many reasons such as the surgical trauma, tourniquet, positioning and indeed the anaesthetic drugs and technique. This has resulted in greater preoperative care by detailed patient history evaluation and examination, patient information and informed consent. Besides the effect of the anaesthetic technique upon the course of the disease, there is also the interaction of drugs administered during anaesthesia and patient medication. Several undiagnosed diseases may be disclosed following a surgical/anaesthetic intervention.

This overview will discuss some anaesthetic considerations with special attention to regional techniques. Many 'conservative' colleagues will be reluctant to consider a locoregional technique when some neurological problems would exist before surgery or delivery. This is mainly because of litigation issues rather than a real contraindication. In addition, recent concern with respect to neural and muscular toxicity of some local anaesthetics has further cast some doubt, even more than in a healthy population, on the benefit of regional anaesthesia.

It should be emphasized that high-risk patients may not infrequently benefit from a locoregional technique. Peripheral nerve blocks may have less respiratory or haemodynamic consequences than neuraxial or general an-

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aesthesia. Obstetric patients are more likely to request regional analgesia or anaesthesia. Therefore the selection of general or locoregional anaesthesia should be made on an individual basis.

Patient information is very important. The problem of neurological complications and relapses or deterioration of the disease should be discussed in advance as these patients are very concerned about this. Many patients are afraid of another needle in the back because they had bad experience with diagnostic punctures. They should also be reassured that motor weakness and sensory block have nothing to do with further progress or relapse of their deficit.

In general, the literature has not changed dramatically during the last decade because it is extremely difficult, if not unethical to perform randomized controlled studies comparing general and regional techniques. Suggestions, with respect to the optimal anaesthetic technique are mostly inspired by anecdotal reports, small sample sizes or animal findings.

Spinal cord injury. Para- and quadriplegia

Spinal cord injury results from trauma, bleeding or a tumoral process and may have consequences on different systems. Bone decalcification, muscle spasms, pressure sores, deep venous thrombosis, thermoregulation problems, renal disease, infections, cardiac rhythm problems and respiratory failure have all been described. In pregnancy preterm labour, pre-eclampsia (sometimes difficult differential diagnoses with autonomic hyperreflexia) and lack of labor progress may all be possible.

Many of the consequences will depend on the level of transection. Initially a paralysis with muscle weakness will predominate while progressively muscles will become spastic.

Lower extremity or abdominal surgery and also delivery may theoretically be performed without anaesthesia if there is sensory loss at the operative site. However autonomic hyperreflexia may occur especially with lesions higher than T7. Skin trauma, distention or examination of hollow viscus such as bladder, bowel or uterus but also distention/contraction of the perineum may cause autonomic hyperreflexia. Afferent stimulation of the adrenal glands will cause catecholamine release. Blood vessels below the lesion are very sensitive to catecholamines because their action is unopposed by descending pathways. The subsequent hypertensive crisis may cause headache, flushing, pupillary dilatation, seizures and intracranial bleeding. Baroreceptors located in the carotis and aortic arch will stimulate the vagal nerve resulting in severe bradycardia.

Depending on the respiratory status, as inspiratory muscles may be in-

volved, general anaesthesia may be the only option, but a deep plane of anaesthesia is required to prevent hyperreflexia, and this may have other consequences such as uterine atony, dysrhythmia etc. If for caesarean section general anaesthesia is considered, a rapid sequence induction is mandatory but it will face the anesthetist with another problem as succinylcholine is contraindicated in paraplegic patients risking a cardiac arrest due to massive potassium release. Fiberoptic intubation may be an alternative (1).

Locoregional anaesthesia such as a neuraxial technique may offer the best haemodynamic stability, but a flexible catheter technique (CSE, CSA or continuous epidural) should be considered rather than a single injection (2-4).

Due to back surgery and muscle spasm the changed anatomy may make performance of neuraxial techniques sometimes very challenging while the level of the block cannot be judged accurately. Sometimes the desired level can not be obtained because of persisting compression of the neural structures or previous surgery (3). Some authors rather prefer isobaric solutions as their spread is more predictable and less dependent on position (5).

Also peripheral nerve blocks have been performed such as interscalene blocks for shoulder surgery in a quadriplegic patient (6).

Timing of regional techniques in relationship to DVT prophylaxis should be considered. Neuraxial administration of opioids alone may not be sufficient. Combinations of local anaesthetics with an opioid allowing a reduction in the concentration of the local anaesthetic seems fine. In case of hypotension ephedrine will be more effective, while adrenaline should be avoided.

It may be recommended to continue postoperative analgesia during the first 24-48 hours.

Another less common complication may be "mass motor reflex" related to handling and positioning of these patients. This is explained by the reflex starting with stimulation of a muscle spindle, which normally causes contraction of one single muscle unit, while in these patients a whole muscle group will respond with a spasm.

Space occupying lesions

Depending on the location of the tumor, general or regional anaesthesia may be indicated.

Due to water and salt retention symptoms may become apparent during surgery or pregnancy. The most important issue may be the degree of intracranial pressure (ICP) increase. General anaesthesia may be beneficial because hyperventilation may enable reductions of ICP, although intubation and the use of succinylcholine (fasciculations) may worsen the preoperative condition. Neuraxial blocks may offer an excellent solution, but several

problems should be considered. The injection of high volumes epidurally may further increase intracranial pressure (7), while accidental taps may have catastrophic consequences, as well due to as cerebellar herniation. It may be subject of debate whether intended spinal anesthesia with small pencilpoint needles might not be a better solution than epidural anaesthesia. Neuraxial analgesia will reduce pain and bearing down (during delivery) which both may increase ICP.

Cerebrovascular disease

This is caused mainly by bleeding from cerebral aneurysms or AV malformations, anticoagulation, while also stroke will fall in this class of problems. Pre-operative or pre-delivery deficit should be documented. The choice of the anaesthetic technique depends mainly on the existence of ICP increase and whether surgical repair has been performed. When surgically corrected, no formal contraindications exist to perform a neuraxial block. With increases of ICP the same precautions and considerations account as for space occupying lesions. When not previously diagnosed or not corrected surgically, the risk of bleeding, more likely during labour or C-section (8) will be very real due to blood pressure increase following endotracheal intubation, pain, pre-eclampsia, valsalva manoeuvres etc. (in fact immediately after the Valsalva due to CSF pressure decrease and increased venous return) all of which may be arguments in favour of regional anaesthesia. However, a decrease in CSF pressure following dural puncture may increase the transmural pressure of the affected vessel, causing subarachnoid haemorrhage. Major intracranial bleeding of an unknown AV-malformation has been reported following an accidental dural tap (9).

The most important issue is to maintain haemodynamic stability. Hypotension may induce vomiting which also will enhance the risks. The use of opioids alone during the first stage of labour has been suggested, as this will not affect haemodynamics, although it may enhance vomiting. On the other hand hypotension during labour will be rare when using low dose combinations. CSA may offer more cardiovascular stability and success than single dose spinal or epidural but a catheter-over-needle technique deserves preference as CSF leakage will be less than with catheter-through-needle equipment.

Perispinal vascular malformations may also exist in certain diseases such as Von Hippel Lindau, Klippel-Trenaunay etc. which should be ruled out by preoperative MRI. However in most case reports, general anaesthesia was selected for uterine surgery or C-section, while labour analgesia has been reported with IV sufentanil and pudendal blocks (10,11).

Traumatic intracranial bleeding may also occur, but once operated re-

gional anaesthesia and even vaginal delivery do not seem to be contraindicated.

The use of transcranial Doppler has been recommended to monitor the cerebral circulation through the middle cerebral artery during C-section under epidural or spinal anaesthesia to prevent further ischaemic damage (12).

Epilepsy

Patients treated for epilepsy should continue this therapy until the time of surgery. After-surgery medication should be given parenterally if oral intake is not desirable. Plasma levels of some substances can be measured accurately. When general anaesthesia would be selected it should be remembered that significant drug interactions may exist between anticonvulsant drugs and substances used during anaesthesia eg. muscle relaxants, hypnotics etc. Phenobarbital may accelerate biotransformation of anaesthetic drugs while diphantoine and carbamazepine may cause resistance to non-depolarizing muscle relaxants. Some epileptogenic anaesthetics should be avoided such as enflurane, ketamine and etomidate. Despite reports of uneventful use of these substances, it may be wise to avoid them because there are several alternatives available. The effects of fentanyl and its analogues, especially in large doses, and propofol have also been subject of debate. Propofol, despite reports of abnormal movements, does not have seizure activity, but depresses EEG at least similar to thiopenthal.

Regional anaesthetic techniques are not contraindicated in these patients. Despite the fact that convulsions may occur with toxic plasmalevels of local anaesthetics, low plasma concentrations may even have potent anticonvulsant effects.

In order to keep the plasmalevels as low as possible, the use of adrenaline may be considered, while appropriate doses and concentrations (of preferably the newer less toxic local anaesthetics) should be injected by incremental doses. Regional anaesthesia/analgesia may offer the additional benefit that hyperventilation (during IPPV or labour) resulting in triggering hypocapnia is avoided. The anaesthetist should be careful with additional sedation especially in patients treated with phenobarbital. Oversedation may cause hypercapnia and acidosis also decreasing the seizure threshold.

It has been suggested that spinal anaesthesia might induce a higher incidence of perioperative seizures (13). It is unclear whether, if true, this is caused by the higher local anaesthetic concentrations in CSF or altered CSF-dynamics (14).

Sclerotic disease

Multiple sclerosis (MS) is a degenerative disease characterized by multiple sites of demyelination of the white matter in the brain and spinal cord, but not in the peripheral nerves. Autonomic dysfunction is equally possible resulting in (orthostatic) hypotension.

When selecting the optimal anaesthetic technique, much will depend on the respiratory function (although mostly the lower trunk is involved first) and interaction with the medication of the patient. Succinylcholine should be avoided (as in any other disease with muscle atrophy). Patients may be resistant to some non-depolarizing relaxants.

The most common fear in the peri-operative period is that exacerbations of the pre-existing deficits may depend on the anaesthetic technique used. It should be emphasized that any stressful condition, fever/hyperpyrexia, infection even without fever, surgery and fatigue may cause exacerbations or relapses. This relapse may occur up to three months following surgery or delivery and may be three times higher than at any other time. Nevertheless pregnancy itself does not seem to affect the course of the disease. During pregnancy symptoms may even ameliorate as with other immunological diseases. Although numbers of patients studied are mostly too low and no controlled studies exist, it has been suggested that general anaesthesia causes less exacerbations, while epidurals and especially spinals do (15) as cases have been reported of an acute onset of previously undiagnosed MS (16). This has been explained by either direct toxicity of the local anesthetic within the CSF or rather high doses of local anaesthetics as with secondary C-section after epidural labour analgesia. Fortunately labours may be shorter thus reducing the amount of local anaesthetics (LA) required. Although the evidence for more relapses with spinal techniques is not strong, epidural seems to be less harmful than spinal anaesthesia lower LA concentrations (25% as compared to spinal) will be measured in the spinal cord white matter (17). In general lower concentrations are recommended than in healthy patients. Hypotension, also more likely with spinal anaesthesia may be resistant to vasopressors while causing further damage due to ischaemia (18). As the blood-brain barrier may be damaged this may be an additional reason why many anaesthetists will avoid spinal anaesthesia. Dural puncture itself does not seem to cause exacerbations. Little is known about possible benefit of opioids, rather than local anaesthetics. It is also recommended not to use adrenaline. Peripheral nerve blocks do not signify any additional problem as only the CNS is involved.

Amyotrophic lateral sclerosis (ALS) involves the anterior horn cells caus-

ing motor neuron disease throughout the central nervous system. As opposed to multiple sclerosis the upper trunk is mostly the first involved which may affect respiratory and swallowing function due to muscle atrophy and weakness. It is controversial whether also with this disease autonomic dysfunction is possible. The respiratory status may determine the choice between neuraxial blocks or general anaesthesia. These patients are usually more sensitive to non-depolarizing muscle relaxants. Little is known about the effect of anaesthesia upon the course of the disease. Also here uneventful epidural anaesthesia has been reported (19) and seems to receive some preference over spinal anaesthesia.

A recent review containing 139 patients with a pre-existing CNS disorder, revealed that there were no patients with new or worsening deficits after the operation performed under neuraxial anaesthesia (20). In this study 40 patients had MS or ALS. However, no subgroup analysis was performed.

Careful preoperative examination is mandatory with documentation of the existing deficit. Possible pros and cons of the different anaesthetic techniques should be discussed with the patient.

Parkinson's disease

This disease is caused by a loss of dopaminergic fibers and dopamine-containing nerve cells in the basal ganglia of the brain. It results in spontaneous movements, rigidity, facial immobility and tremor. These symptoms are secondary to diminished inhibition of the extrapyramidal motor system. Levodopa, the precursor of dopamine, is still the most effective drug despite the search for alternatives such as drugs preventing dopamine degradation. To lower the dose of levodopa it is combined with an inhibitor of decarboxylase, as this enzyme is also present in the circulation. However this treatment, still the most universally given, may further enhance the risk of orthostatic hypotension already present in some stages of the disease. Other possible drugs in Parkinson's disease are MAO-inhibitors, other anti-depressants, anti-spastic drugs etc.

Anaesthetic intervention may be necessary eg. for the newer neurosurgical therapeutic approaches. The most important considerations during anaesthesia are the mostly older ages of these patients and the interaction with (mostly multiple) anti-Parkinson medications (21). Interruption of levodopa therapy for more than 6 hours may cause muscle rigidity. Neuroleptic substances and metoclopramide should be avoided because they may antagonize the effects of dopamine. Autonomic dysfunction is very common and may cause hypotension, gastric paresis and excessive salivation. Although the information about regional anaesthesia is meagre, it offers the advan-

tage that oral medication can be continued. However, neuraxial techniques, especially spinal anaesthesia may induce significant hypotension due to the combination of the disease and its treatment, both already tending to cause vasodilation.

Other central nervous system diseases

The incidences of these diseases are mostly rather low, which does not allow to draw conclusions with respect to the type of anaesthesia to be selected. Patients with poliomyelitis, Friedreich's ataxia, Huntington chorea, syringomyely etc. may require a surgical intervention and spinal/epidural anaesthetic techniques have been reported to be uneventful (22-24). General anaesthesia may be desirable in case of dementia or uncontrolled movements but the risk of aspiration should also be considered.

Nowadays HIV+ patients are more frequently encountered in operative theatres, and although they may suffer central nervous system problems, regional anaesthesia is surely not contraindicated. There has been concern about the possibility to spread the virus in the CSF when performing a spinal technique but evidence for this is very poor (25). The choice between regional and general anaesthesia will mainly depend on the thrombocyte count and the mental status and cooperation of the patient.

Chronic back problems and previous spine surgery

Pre-existing back pain receives increasing attention with respect to possible worsening of the condition following neuraxial techniques. Nevertheless, epidural infiltrations with corticosteroids have been used for decades to treat chronic back problems.

Based upon several cases in the literature, some of them already reported 30 years ago (26-30) I have growing concerns about performing neuraxial techniques in patients with spinal stenosis. In Belgium, claims are known of neurological deficit (cauda equina, motor weakness, sensory deficit etc.) following neuraxial blocks performed in patients in whom, though relatively symptomless, the diagnosis of spinal stenosis was evidenced after the appearance of postoperative problems. It is unclear whether this is caused by the spinal injection as part of a CSE technique, the choice of the drugs or the volume of administration. Although most claims have not found the anaesthetist to be blamed, a relationship between the anaesthetic technique and the problems experienced by the patient can hardly be ignored.

Pregnant patients may suffer more back and radicular pain than a non-pregnant population, even persisting for several months after delivery. Differing orthostatics or direct compression of the lumbar plexus are the main

reasons but a herniated disc may equally be possible necessitating pre- or post-delivery surgery. Decision making is not easy, when these patients prefer a regional technique but based upon personal experience I agree with a rather reassuring recent review (31), which could not find occurrence of new or progressive deficit (see below).

If patients have been operated for back problems, this may be very challenging for the anaesthetist although much will depend on the extent of surgery. Many handbooks still consider back surgery as a contraindication to the use of regional anaesthesia. In addition, the patient may be reluctant as well to undergo epidural or spinal anaesthesia fearing exacerbation of the pre-existing problems. Extensive surgery may not enable regional anaesthesia from a technical point of view. When puncturing the back in the vicinity of previously performed surgery, there is an enhanced risk of dural tap although according to personal experience PDPH will be less likely to occur. When an EBP is required then similar technical difficulties should be anticipated.

Besides the risk of an inadvertent dural tap, patients having undergone previous back surgery are at a higher risk of technical problems, traumatic needle placement, incomplete or patchy blocks and maldistribution of LA, due to adhesions, causing more toxic concentrations in certain areas. Especially epidural anaesthesia may be a particular challenge in operated patients. After Harrington rod instrumentation in close to 50% of patients placement of an epidural catheter may not be successful on the first attempt, while many will require larger doses or complain of incomplete or patchy blocks (32-34). However, even multiple punctures may still offer fair success rates. Especially when the fusion terminates above L3-L4, the success rate of an epidural may be increased.

A french group found 18% overall failure rate (9% technical and 9% analgesic failure) with epidurals in 31 parturients with previous spine surgery (35).

A spinal technique (CSE, CSA, SDS) may receive some preference in these patients. In a prospective study of 30 delivering women with severe untreated scoliosis, corrected scoliosis or lumbar-sacral fusion, 19 of these received CSA with only one case of post dural puncture headache (36). A SDS may cause a lower incidence of paraesthesias as compared to catheter techniques. Paraesthesias are not infrequently brought in relationship with possible postoperative neurological deficit (37).

A recent review has focused on the risk of neuraxial blocks in patients suffering spinal stenosis or lumbar disc disease with some of them having undergone discectomy or laminectomy (31). In this report success-rates did not differ. One percent experienced new or progressive deficit although the

majority was related to the surgical trauma or tourniquet. Although this review may suggest that patients suffering lumbar disc disease or spinal stenosis may receive a neuraxial block whether or not they have been operated, again no subgroup analysis was done for the spinal stenosis group for which I would recommend prudence.

Finally, patients with implanted pumps or spinal cord stimulators have been subject of concern for neuraxial anaesthetic techniques because of the risk of damaging the implant, infection, and unknown effects of the injected drugs or blood (in case of a blood patch). Nevertheless neuraxial blockade has been reported to be uneventful (38,39).

Peripheral neurological deficit

Peripheral nerve blocks may theoretically cause further harm, if deficit is already existing. This may be explained by direct needle or catheter trauma, local anaesthetic toxicity, neural ischaemia or a combinations of factors. In a recent review in the MAYO clinic dealing with 360 patients undergoing ulnar nerve transposition, 100 patients received an axillary block (40). In both the general and axillary group 6% of the patients experienced new or worsening deficit. In the axillary group all of them received bupivacaine which was considered as an independent risk factor compared with other local anesthetics whereas ulnar paraesthesia or motor response were not. The authors concluded that the anaesthetic technique in this type of disease (mostly mononeuropathy) did not affect neurological outcome but that this may not necessarily account for toxic or metabolic neuropathy or in the presence of other neurological disease.

Diabetes will cause a high incidence of peripheral neuropathy which is not always noticed before surgery. Not unfrequently anaesthesia is blamed for neurological deficit such as sensory loss or pain (may follow wrong positioning) which might be caused by other reasons such as delivery with passage of the foetal head through the pelvis. Oedema of the nerves may be directly related to hyperglycemia. Microangiopathy may further lead to ischaemia of the oedematous nerve, but also decrease the uptake of the local anaesthetic at the injection site resulting in prolonged exposure. It is unclear whether and which local anaesthetics may further enhance oedema and ischaemia of the nerves. As also the spinal cord is involved, neuraxial blocks may theoretically predispose diabetics to injury.

Autonomic neuropathy on the other hand, may cause tachycardia, orthostatic hypotension, gastric paresis and micturition problems. Neuraxial blocks will cause more hypotension. As a consequence of both peripheral and autonomic neuropathy diabetics should receive lower doses and concentra-

tions of local anaesthetics than non-diabetics. However, a recent review of a large number of patients suffering peripheral neuropathy could not find an increased risk of neurological complications following neuraxial blockade (41).

Although avoidance of adrenaline may account for practically all patients suffering neurological disease, the diabetic patient seems to be the most vulnerable to the use of vasoconstrictors.

Several studies have demonstrated that peripheral blocks, more particularly popliteal blocks, may require more needle passes to observe the desired motor response upon nerve stimulation, but afterwards result in higher success rates and longerlasting anaesthesia without further neurological complications (42,43). Changing the current and/or its duration but also the use of Doppler technology may increase the success rate of nerve blocks in these patients.

Guillain-Barré syndrome also known as polyradiculoneuritis results from a viral infection and causes acute or subacute muscle weakness progressing cephalad. Although full or good recovery occurs in 85% of the cases, in 3-5% chronic or recurrent neuropathy may persist. Autonomic dysfunction may cause hypotension but following intubation hyperreflexia may be possible as well. Potassium release after succinylcholine administration may be at risk even some time after recovery. The effect of nondepolarising muscle relaxants is unpredictable and is related to the stage (denervation versus reinnervation) of the disease.

Experience with regional anaesthesia is poor, but epidural opioids can be administered safely for the management of pain and unpleasant sensory disturbances.

Myasthenia gravis

Myasthenia will be caused by destruction of acetylcholine receptors. Treatment consists of anticholinesterase drugs, but sometimes patients need to be treated with corticosteroids, immunosuppressive substances, plasmapheresis or thymectomy. Thymectomy through sternotomy may be very challenging, as in the postoperative period it may further compromise respiratory function. The risk of postoperative ventilatory insufficiency is enhanced when myasthenia exists for more than 6 years, presence of previous respiratory problems, pyridostigmine doses >750mg/day or a preoperative vital capacity <2.9L. Rapid deterioration in the postoperative period is possible as a result of stress, fatigue, infection or anti-cholinesterase overdose.

General anaesthesia may be the best option when respiratory function is severely affected. In the '70 general anaesthesia was also recommended

for C-section (44). Succinylcholine may result in an unpredictable duration depending on anti-cholinesterase therapy which may prolong its activity or cause a dual block. Non-depolarising muscle relaxants should be administered in very weak doses preferentially while monitoring neuromuscular function. Increased sensitivity is present even in cured and symptomless patients. In fact, muscle relaxants should be avoided whenever possible.

Nowadays, locoregional anaesthesia may be an interesting alternative, but weak concentrations of local anaesthetics causing the least motor impairment should be selected. Also for labour pain epidural analgesia may be beneficial, because it will decrease stress and fatigue. Using spinal opioids alone for the first stage may be considered. Perineal anaesthesia allows forceps delivery to shorten the second stage and decrease fatigue. A report of 10 cases has shown that both labour analgesia and C-section anaesthesia may be safely performed with an epidural (45). A single dose spinal may affect respiratory muscles more deeply and unpredictably than an epidural or low-dose CSE technique.

Myopathy

Mainly muscular dystrophy and myotonic disorders will be discussed in this section.

Among **muscular dystrophies** Duchenne's disease is the most severe form whereas Becker muscular dystrophy occurs later and has a slower evolution. Both diseases are X-chromosome linked, result from either no or dysfunctional dystrophin and cause elevated levels of creatine kinase (CK). Skeletal muscle degeneration and atrophy are caused by cell damage. Rhabdomyolysis is also associated with Duchenne's disease. Death results from respiratory insufficiency, pneumonia or heart failure as also smooth and cardiac muscles may be damaged although the severity of skeletal muscle degeneration does not correlate with the degree of cardiac involvement.

When anaesthesia is necessary the most significant complication may be the unexpected effect of anaesthetic drugs upon myocardial function. Sudden perioperative cardiac arrest is known for several decades but still occurs in the 21st century (46,47). Duchenne's myopathy may remain undiagnosed until an anaesthetic mishap occurs mostly following the induction with succinylcholine (47,48). The risk of aspiration of gastric content should be considered, which accounts for all myopathies, but succinylcholine should be avoided to prevent hyperkalemia. The onset of rocuronium on the other hand, when used to enable rapid sequence intubation has been found to be prolonged by a factor 2 as compared to controls (49). The onset of mivacurium does not seem to be prolonged (50). However the duration of

action of rocuronium and mivacurium has been found to be significantly prolonged (49,50). Succinylcholine may also cause malignant hyperthermia (MH). Although several reports of MH in Duchenne's dystrophy exists (51), the association between both diseases is not commonly accepted.

Many types of **myotonia** exist. It is a persistent contraction of skeletal muscle after voluntary contraction or stimulation. Mostly stiffness decreases with exercise as in Thomsen's disease (myotonia congenita), except for paramyotonia congenita. Myotonic dystrophy, better known as Steinert's disease, is the only disorder in the myotonic group also causing extramuscular effects including cataract, gastrointestinal, endocrine and cardiac (mainly valvular and conduction) abnormalities.

Restrictive lung disease may be caused by weakness of respiratory muscles. Less effective cough may cause pneumonia. Patients may require prolonged postoperative ventilation. Myotonia may be induced during surgery and anaesthesia by cold, shivering, drugs such as neostigmine (although subject to debate), propofol or succinylcholine, manipulation, nerve stimulators, and electrocautery. Myotonic contractures will not be abolished by regional blocks, deep anaesthesia or muscle relaxants and may compromise intubation and/or ventilation. Differential diagnosis with MH may be difficult although there may not be a reason to suggest an association between both.

With respect to regional anaesthesia in myopathy, little is known unless that it may be beneficial as compared to general anaesthesia and that it will not treat or prevent the occurrence of myotonic contractures. This does not preclude the use of spinal and epidural anaesthesia in pregnant and non-pregnant patients but care should be taken (as with general anaesthesia) not to allow body temperature to decrease too much. Sympathicolysis may enhance loss of body heat and shivering while this may be more pronounced during labour. Uneventful epidural and CSE anaesthesia has been reported for C-section in patients with muscular dystrophy or being a carrier (52,53).

In all myopathies there seems to be an enhanced risk of scoliosis for which surgical correction may be mandatory to improve the pulmonary function. This may cause technical difficulty. General anaesthesia combined with CSA for postoperative analgesia with morphine has been reported in a scoliotic patient with Duchenne's disease (54). Also for myotonic dystrophy the use of epidural local anaesthetics and/or opioids has been recommended to improve pulmonary function (55). However, being very sensitive to opioid induced respiratory depressant effects, care should be taken in these cases.

Finally, with recent studies on myonecrosis following bupivacaine, the fu-

ture needs to determine which local anaesthetics may be safer than others in these cases (56).

Conclusion

Pre-existing neuromuscular disease may be a challenge to the anaesthetist. Probably the experience with general anaesthesia is much larger than with regional techniques. The effect of anaesthesia upon the disease needs to be considered, as well as the effect of the disease and patient chronic medication upon the course of anaesthesia.

As general anaesthesia requires more manipulations and drugs to be administered, it seems logical that this may cause more significant effects than regional anaesthesia.

Nevertheless, when faced with patients with preoperative neurological disease, many anaesthetists will consider this as a contraindication for regional anaesthesia despite a lack of controlled studies. Based upon anecdotal reports the medicolegal issue continues. A good preoperative examination remains mandatory while patients should be informed about technical difficulties, possible relapses and/or progression associated with the combination of stress, surgery and anaesthesia.

Patients may suffer further deficit by the anaesthetic technique selected (multiple attempts, catheters, needle trauma, vasopressor, toxicity etc.). It should be recommended that paraesthesia, adrenaline and high concentrations of local anaesthetics should be avoided at all times. Some diseases may benefit from epidural/peripheral anaesthesia (multiple sclerosis, epilepsy, space occupying lesions, myasthenia) while for others a spinal technique may be the best choice (spine surgery, opioid-alone analgesia).

Special attention should be paid to patients with spinal stenosis despite recent reports in favour of regional anaesthetic techniques.

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