Case report: Successful labour epidural analgesia in a patient with spinocerebellar ataxia

[Analgésie péridurale pour travail obstétrical réussie chez une patiente souffrant d'ataxie spinocérébelleuse]

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Purpose: To report the favourable outcome associated with epidural analgesia in a parturient with spinocerebellar ataxia (SCA).

Clinical Features: A 34-yr-old patient, G2 P0, presented at term with a history of SCA since the age of 22 characterized by slurred speech, balance and gait disturbances, diplopia and nystagmus. A magnetic resonance imaging of the brain at the age of 27 showed cerebellar degeneration. Neurological examination revealed an unsteady, wide-based gait, nystagmus, mild dysarthria, moderate finger to nose ataxia, absent reflexes in all upper and lower limbs, sensory loss to vibration and temperature discrimination up to the level of both knees, and normal motor strength. The patient presented for induction of labour at 40 weeks and requested epidural analgesia, which was performed in the usual manner. Following a negative test dose of 3 mL of 2% lidocaine, a loading dose of 10 mL of 0.125% bupivacaine was administered, and maintenance of analgesia was achieved with a mixture of bupivacaine 0.0625% and fentanyl 2 $\mu g \cdot mL^{-1}$. The patient required standard doses of the epidural mixture, and experienced effective analgesia for labour and delivery. Her recovery was uneventful and no subsequent neurological deficit was detected up to two years after delivery.

Conclusions: Neurological disorders may contraindicate regional anesthesia, and the decision to proceed with a regional technique should be based on the pathophysiology and severity of each particular case. Uneventful epidural analgesia was provided to a parturient with SCA, with no long term effects detected up to two years after delivery.

CAN J ANESTH 2007 / 54: 6 / pp 467–470

Objectif *: Rendre compte du résultat favorable associé à une analgésie péridurale chez une parturiente souffrant d'ataxie spinocérébelleuse (SCA).*

Éléments cliniques *: Une patiente de 34 ans, G2 P0, arrivée à terme, s'est présentée avec des antécédents de SCA depuis l'enfance, caractérisés par un parler inarticulé, des troubles de l'équilibre et de la marche, de la diplopie et un nystagmus. Une imagerie par résonnance magnétique (MRI) du cerveau à l'âge de 27 ans avait montré une dégénérescence cérébelleuse. L'examen neurologique a révélé une marche peu assurée et en abduction, un nystagmus, une dysarthrie légère, une ataxie modérée lors du test doigt-nez, des réflexes absents aux membres supérieurs et inférieurs, une perte sensorielle aux vibrations et à la discrimination de température jusqu'au niveau des deux genoux, et une force motrice normale. La patiente s'est présentée pour l'induction du travail obstétrical à 40 semaines et a demandé une analgésie péridurale, qui a été effectuée de la manière habituelle. Suite à une dose test négative de 3 mL de lidocaïne 2 %, une dose de charge de 10 mL de bupivacaïne 0,125 % a été administrée, et l'analgésie a été maintenue par un mélange de bupivacaïne 0,0625 % et de fentanyl 2 $\mu g \cdot mL^{-1}$. La patiente a requis des doses habituelles du mélange péridural, et a reçu une analgésie efficace pour le travail obstétrical et l'accouchement. Son rétablissement s'est fait sans incident et aucune déficience neurologique consécutive n'a été détectée dans les deux années suivant l'accouchement.*

Conclusion *: Les troubles neurologiques peuvent contrindiquer une anesthésie régionale, et la décision d'avoir recours à une technique régionale doit se baser sur la pathophysiologie et la gravité de chaque cas en particulier. Une analgésie péridurale sans complications a été fournie à une patiente souffrant de SCA, et aucun effet à long terme n'a été détecté durant les deux années suivant l'accouchement.*

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Conflicts of interest: None.
Accepted for publication February 8, 2007.
Revision accepted March 1, 2007.

SPINOCEREBELLAR ataxias (SCAs) are neurodegenerative disorders characterized by progressive, unremitting ataxia due to involvement of the cerebellum, brain stem, basal ganglia, pyramidal tract, somatosensory spinal pathways, optic nerve and, occasionally, peripheral nerves. Cerebellar ataxia represents the main clinical feature, and the most consistently reported clinical manifestations include lack of coordination while performing voluntary motor activities, unsteady gait and clumsiness. Other manifestations may include dysarthria, dysphagia, facial palsy, hypotonia, tremors, weakness, nystagmus, diplopia, and/or retinal degeneration.[1,2] Spinocerebellar ataxia may be inherited as an autosomal dominant or autosomal recessive pattern, or may also occur by sporadic point mutation. Based on clinical features and genetic mapping, SCAs are divided into multiple subtypes, the majority of which have been assigned specific chromosomal loci.[3] The socioeconomic burden of SCA results from marked impairment in functional status and activities of daily life. There is no efficacious treatment currently available for many of these disorders other than supportive care.[4] There are no available data describing the course of SCA in pregnancy, and the potential effects of regional anesthesia on SCA remain unknown. Based on a literature review, we found this to be the first description of anesthetic management in a parturient with SCA. Written consent for publication of this report was obtained from the patient.

Case report

A 34-yr-old, G2 P0 patient was first seen at the high-risk obstetric anesthesia clinic at 35 weeks gestation. She had a history of SCA, starting at the age of 22, with slurred speech, and balance and gait disturbances. Her condition followed a slowly progressive course with the development of diplopia and nystagmus. She started to use a walker at age 26. A magnetic resonance examination of the brain at age 27 showed cerebellar degeneration. The patient denied any history of cardiovascular, respiratory, bowel, or bladder involvement. Repeated genetic testing for different forms of SCA, including subtypes I, II, III, IV, V, VII and VIII, as well as a test for Friedreich's ataxia, neither identified a specific gene for SCA nor revealed any carrier state of a mutant gene for SCA. Her family history was negative for SCA or any other neurological disorders. She had had two uneventful general anesthetics; at age 22 for excision of a right breast fibroadenoma, and at age 33 for dilation and curettage for termination of a pregnancy at 14 weeks due to the antenatal diagnosis of Trisomy 21. Also, she had had recent foot surgery under a peripheral nerve block, as a neuraxial block was denied because of her history of SCA. During the current pregnancy, the patient developed mild gestational diabetes at 27 weeks, which was controlled by diet. She was confined to a wheelchair throughout pregnancy because of fear of falling, and worsening symptoms of diplopia and nystagmus. Her obstetric plan was for vaginal delivery.

On physical examination, she was 163 cm in height and weighed 68 kg. Her vital signs were within normal limits. Airway, cardiopulmonary and lumbar spine examinations were unremarkable. Neurological examination revealed a downward nystagmus to the left, and lateral gaze evoked nystagmus while looking to the right. Her speech was mildly dysarthritic with no evidence of tongue fasciculations. She had moderate finger to nose ataxia. Her reflexes were absent in all upper and lower limbs, and she had a sensory loss to vibration and temperature to the level of both knees. Her motor strength was normal. She presented with an unsteady, wide-based gait.

A review of the literature and a consultation with neurology was undertaken by the attending anesthesiologists in the days following. The consensus of the multidisciplinary team consisting of the neurologist, obstetrician and anesthesiologist was for regional anesthesia during labour and/or delivery. The findings were explained to the patient, who chose to receive regional anesthesia based on the information that, considering the pathophysiology of the disease process, no absolute contraindication existed for regional techniques.

The patient presented for induction of labour at 40 weeks, and requested epidural analgesia. At the stage of cervical dilation of 2 cm, an epidural catheter was inserted uneventfully, using a standard technique, at the L4–5 interspace. Ten minutes after a negative test dose of 3 mL of 2% lidocaine, a loading dose of 10 mL of 0.125% bupivacaine was administered in two divided aliquots, five minutes apart. Labour analgesia was maintained with patient controlled epidural analgesia (PCEA), using a mixture of bupivacaine 0.0625% plus fentanyl 2 $\mu g \cdot mL^{-1}$. Patient controlled epidural analgesia was initiated with a bolus dose of 5 mL, basal infusion rate of 10 $mL \cdot hr^{-1}$, ten-minute lockout interval, and a 40-mL one-hour limit. Following the loading dose, a T8 sensory block to ice was established bilaterally, and remained stable as per subsequent assessments. No changes in maternal vital signs or fetal heart rate were reported throughout labour. The patient was satisfied with the epidural analgesia, and did not attempt to walk during labour. Patient controlled epidural analgesia continued for

about six hours, until the spontaneous vaginal delivery of a healthy male baby with Apgar scores of 9 and 9 at one and five minutes respectively. A total of 50 mg of bupivacaine was administered over six hours, including the loading dose. One hour after delivery and discontinuation of PCEA, the patient had a bilateral T10 sensory block to ice, and a motor block of 4 on the left and 2 on the right leg according to the modified Bromage scale.[5] Complete resolution of the sensory and motor block occurred within four hours postpartum. The patient was followed daily for the first three postpartum days, during which her neurological assessment remained unchanged from the basal condition.

The patient was followed up through phone interviews. No subsequent neurological deficit has been detected up to two years after delivery, as per the patient's report. This is in accordance with the slowly progressive natural course of this condition. The only new information revealed was the identification of a new gene for ataxia, which established her diagnosis as ataxia with oculomotor apraxia type 2.[6]

Discussion

Although cerebellar involvement is the hallmark in the majority of SCA subtypes, peripheral neuronal involvement has been reported as well, suggesting that the disease pathogenesis is not solely confined to the central nervous system.[1,2] It may take the form of dying-back axonal neuropathy or neuronopathy, with degenerative changes and reduction in the number of neurons in the anterior horns and/or dorsal root ganglia.[7] Although electrophysiological abnormalities are not always reported in patients with peripheral nerve involvement, some patients with electrophysiologically confirmed peripheral nerve involvement do not have any accompanying clinical manifestations, suggesting that the peripheral nerve involvement is often mild and subclinical. The slow progression of disease may also contribute to this observation.[7,8]

The patient described in this report was greatly concerned about the possibility of having an epidural for labour analgesia, particularly because neuraxial anesthesia was denied for her previous foot surgery. There is no evidence in the literature to suggest that the pathologic process may be exacerbated by epidural anesthesia or that the peripheral nerves of patients with SCA may be more sensitive to local anesthetics. On the other hand, because of relatively scant data in the literature, a potential risk of these complications cannot be excluded. Nevertheless, Timmann *et al.*[4] observed improvement in some of the symptoms of SCA type 6, mainly dysarthria, ataxia and gait equilibrium, after *iv* administration of lidocaine. This may be related to calcium channel blocking properties of local anesthetics. Motor weakness caused by motor neuron involvement in SCA represents an anesthetic challenge, due to the anticipated decrease in muscle strength with regional anesthesia. However, motor strength was normal in this patient, which is not uncommon with some subtypes of SCA.[9] Before epidural analgesia was considered, all pre-existing neurological deficits were carefully documented in consultation with the patient's neurologist. We elected to use our standard epidural mixture, a combination of a low concentration local anesthetic solution with opioid. This mixture allows the reduction of the local anesthetic concentration, and thus tends to minimize the potential for direct neurotoxicity and to preserve muscle function to a greater extent than local anesthetics alone.[10,11] Our patient required standard doses of local anesthetic and had effective analgesia throughout labour and delivery.

Successful outcomes with regional anesthesia have been reported in a wide variety of neurological disorders for both obstetric and non-obstetric patients, without subsequent neurological complications.[12,13] Tsen *et al.*[10] reported the successful administration of labour epidural analgesia in a patient with olivopontocerebellar degeneration, a neurological disorder with pathologic and clinical features similar to those of SCA.[1] Also, successful spinal[14] and epidural[15] anesthesia have been reported in patients with Friedreich's ataxia, a combined upper and lower motor neuron disease with predominant cerebellar signs.

The use of regional anesthesia in patients with demyelinating disorders, such as multiple sclerosis (MS), has been controversial. In these pathologic entities, epidural anesthesia appears to be safer than spinal anesthesia, as the latter increases the potential for neurotoxicity through exposing unprotected nerve tissue to direct contact with a higher concentration of local anesthetic.[11] The efficacy of labour epidural analgesia in parturients with MS has been described previously,[13,16] with no apparent increase in the incidence of postpartum relapse; however, relapses were more commonly related to the use of bupivacaine in concentrations greater than 0.25%.[13] Moreover, relapses in patients with MS cannot be easily attributed to any particular anesthetic technique, but they do seem to be related to stress and postoperative infection and/or fever.[11] Spinocerebellar ataxia, unlike these demyelinating and inflammatory conditions, is a slowly progressive degenerative disease that is related to neuronal/axonal death, and does not exhibit a relapse pattern.[2,7]

General anesthesia, when considered for operative delivery, may not represent a superior alternative to regional anesthesia in patients with ataxia. Moreover, general anesthesia may pose additional risks, particularly the risk of aspiration, because dysphagia is a commonly reported manifestation, often resulting in repeated choking and regurgitation spells.[17] Previously described in patients with Huntington's chorea and Friedreich's ataxia, the exaggerated response to barbiturates, and both depolarizing and non-depolarizing muscle relaxants, may also exist in patients with SCA, thus increasing the potential for prolonged mechanical ventilation.[18–20]

In conclusion, we report the effective use of labour epidural analgesia in a parturient with SCA, with no residual impact on the patient's neurological status.

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