Anesthetic Management of a Patient with Dystonia Musculorum Deformans

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A 20-year-old Caucasian woman has dystonia musculorum deformans, a rare, inherited disease of the central nervous system characterized by involuntary movements and torsion spasms. She has opisthotonus and hyperextension of the neck resulting from prolonged muscular hypertonicity. The onset of this disease is usually between the ages of 5 and 15 years. The diagnosis is clinical; the electromyogram is not diagnostic.

These patients have been treated, in the past, with various drugs such as haloperidol, levodopa, and carbamazepine,¹ as well as by basal-ganglia surgery.² Steen² has reported on the anesthetic management of these patients for basal-ganglia surgery. He suggests that the fixed cervical spine in the adult may make it impossible to maintain the airway once anesthesia is induced; therefore, awake intubation may be indicated.

Recent developments in neurosurgery have shown that anterior cerebellar electrical stimulation inhibits postural reflexes in cats and man. In the latter, Cooper² has used cerebellar stimulation for the relief of spasticity in stroke, cerebral palsy, and dystonia. This is a case report of a patient scheduled for insertion of anterior cerebellar stimulators through a suboccipital craniectomy for control of intractable spasticity.

REPORT OF A CASE

In infancy the patient developed weakness of her left foot, which was corrected when she was 11 years old by a tendon reinsertion. Thereafter, athetoid movements appeared in her left foot and hand. A rapid increase in her symptoms occurred during the year prior to the present operation. Significant developments included neck retractions leading to hyperextension. Only with great effort could she flex her neck for 30 to 45 seconds (fig. 1). Neck retractions were so severe that the patient could not feed herself. She was unable to walk without assistance, and finally was confined to a wheelchair prior to hospitalization.

On physical examination, the head and back could be somewhat straightened out with pressure, and there did not appear to be excessive shortening or fibrosis of the neck muscles. The mouth and jaw appeared normal and could be opened wide.

The patient was premedicated with diazepam, 10 mg, and atropine, 0.4 mg, and arrived in the operating room calm and alert. Anesthesia was induced with diazepam, 15 mg, and thiopental, 200 mg, iv. These drugs made the patient unresponsive to verbal commands but did not relax her. She maintained an unobstructed airway and adequate spontaneous ventilation. Enflurane in oxygen was

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then administered by mask, increasing the concentration to 4 vol per cent over a 15-minute period. The patient gradually became less spastic and could be straightened to a neutral posture, although the posterior muscles of the neck remained tight. Ventilation could be easily assisted with bag and mask. Pancuronium, 4 mg, iv, was administered to facilitate endotracheal intubation. The patient was placed in the sitting position for the operative procedure. The neck could be flexed sufficiently for surgical exposure. The four-hour anesthetic course was uneventful. Anesthesia was maintained with approximately 1.5 vol per cent inspired enflurane with 60 per cent nitrous oxide at a total flow rate of 3 l/min. Ventilation was controlled and blood gases remained within normal limits.

The trachea was extubated in the operating room, where the patient was able to carry on a simple conversation and had no apparent neurologic sequelae. She initially maintained a normal position, which gradually reverted to her original posture during the first four hours after operation.

On the third postoperative day, the cerebellar stimulators were activated, with no immediate change in posture, but a sensation of relaxation and cessation of twitching movements of the mouth and hands. Over the ensuing three weeks the neck spasms were reduced in number and intensity, allowing the patient to lie on her back. She was able to feed herself and sleep well for the first time in a year. She was discharged eight weeks postoperatively, while the opisthotonus continued to improve.

DISCUSSION

Enflurane was chosen as the primary anesthetic because of its muscle-relaxing action. Patients with dystonia are known to relax with sleep; therefore, we decided to proceed with deep anesthesia and muscle relaxation and intubate the trachea in this way rather than resorting to a tracheostomy. In dystonia, myoclonus deformans neither depolarizing nor nondepolarizing relaxants are contraindicated after the anesthetist demonstrates that the patient can be ventilated by bag and mask in spite of the abnormal posture. Muscle relaxants will not increase the spasticity.

The management of this case of dystonia, myoclonus deformans presents clinical evidence that enflurane can produce conditions favorable for endotracheal intubation in a patient with severe postural deformities. Muscle relaxants are not contraindicated in dystonia, myoclonus deformans; however, profound muscle relaxation can be produced by deep enflurane anesthesia.

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REFERENCES


Obstetrical Anesthesia

ANESTHESIA FOR POSTPARTUM TUBAL LIGATION Twenty-six puerperal patients received 15 mg bupivacaine (Nisentil) and 15 mg diazepam (Valium) intravenously in addition to 100 mg bupivacaine intradurally and 400 mg lidocaine intraperitoneally for postpartum tubal ligation. This volume of local anesthetic provided satisfactory analgesia, with blood levels of lidocaine well below those at which toxicity occurs. Patient and physician acceptance of the technique was good. No complications were recorded. (Cruikshank D. P., Laube D. W., and De Backer L. J.: Intraperitoneal Lidocaine Anesthesia for Postpartum Tubal Ligation. Obstet Gynecol 42:127, 1973.)