Clinical Reports

Refractory dystonia during propofol anaesthesia in a patient with torticollis-dystonia disorder

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***Purpose**: To report a case of refractory dystonia under propofol anaesthesia in a patient with Torticollis-Dystonia disorder.*
***Clinical features**: A 38-yr-old man presented for an MRI scan for investigation of a Torticollis-Dystonia disorder. There was a biphasic response to propofol with complete amelioration of the torticollis and limb dystonia initially with subsequent recurrence under deep propofol anaesthesia. Coadministration of midazolam, diazepam, and thiopentone were not successful in abolishing the recurrent dystonia.*
***Conclusions**: Propofol should preferably be avoided in patients with torticollis and dystonias. Where complete control of movements is required, it may be necessary to consider general endotracheal anaesthesia with muscle relaxants.*

***Objectif**: Rapporter un cas de dystonie réfractaire chez un patient souffrant de torticolis et de dystonie.*
***Éléments cliniques**: Un patient s'est présenté en IRM pour investigation d'un torticolis associé à une dystonie. La réponse au propofol était biphasique et composée initialement de la disparition complète du torticolis et de la dystonie aux membres avec récurrence subséquente après l'approfondissement de l'anesthésie au propofol. L'administration conjointe de midazolam, de diazépam et de thiopentone n'a pas réussi à abolire la récidive de la dystonie.*
***Conclusion**: Le propofol ne devrait pas être utilisé en présence de torticolis et de dystonie. Là où un contrôle absolu des mouvements est requis, il faut peut-être considérer une anesthésie générale endotrachéale avec relaxation musculaire.*

Key words
ANAESTHETICS, INTRAVENOUS: propofol, thiopentone;
BRAIN: extrapyramidal system symptoms;
COMPLICATIONS: hyperreflexia;
MUSCLE: rigidity.

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A 38-yr-old man presented for an MRI examination of his head for evaluation of spasmodic torticollis associated with hemidystonia. He gave a history of developing spasmodic torticollis one year after a rodeo accident in 1993 during which he sustained a right parietal skull fracture and loss of consciousness. The patient underwent operative repair of the fracture under general anaesthesia. The spasmodic torticollis had both tonic and phasic characteristics. The abnormal postures disappeared completely during sleep. On examination his dystonia extended beyond the range of spasmodic torticollis and involved the shoulder girdle and trunk on the left side. The phasic movements (5–8 per minute) of the head and shoulders resembled choreic movements and had a component of tremor. Besides the hemidystonia, his neurological abnormalities included pyramidal signs (hyperreflexia and plantar extension) in his lower limbs and mild, bilateral, symmetrical high-arched feet and hammer toes. The patient was otherwise healthy, was not receiving medications and weighed 110 kg.

In the MRI suite an intravenous infusion of normal saline was commenced and routine monitors (pulse oximetry, ECG and non-invasive blood pressure) were applied. Oxygen was delivered by an aerosol face mask. Premedication consisted of 2 mg midazolam *iv*. After an initial bolus of 0.5 $mg \cdot kg^{-1}$ propofol, an infusion was commenced with a BARD® infusion pump (located

outside the MRI room) and the dose was titrated between 50–100 $\mu g \cdot kg^{-1} \cdot min^{-1}$ to achieve sedation and abolish aberrant spasmodic movements. The weight dial of the pump was set at its maximum of 100 kg. After 45 min of anaesthesia the patient started developing phasic movements of the shoulder girdle (approximately 3–5 every five minutes). The infusion was running well and there were no mechanical problems with the pump. The propofol infusion rate was increased to 160 $\mu g \cdot kg^{-1} \cdot min^{-1}$ after a bolus of 100 $\mu g \cdot kg^{-1}$. It was noticed that some of the phasic movements were closely associated with inflation of the automatic blood pressure cuff. Satisfactory control of the movement disorder was obtained for a further 45 min. Subsequently, the dystonic movements recurred with increased amplitude. Because the patient had already received premedication with midazolam a further small dose of 1 mg midazolam *iv* was administered without benefit. Consideration was given to using a muscle relaxant with tracheal intubation for the rest of the procedure but it was felt that deeper anaesthesia with propofol may be successful. Therefore, a laryngeal mask airway was inserted after a further bolus of 200 $\mu g \cdot kg^{-1}$ propofol. The patient continued to breathe oxygen via the laryngeal mask and nitrous oxide was not used because it is not available in the MRI suite. The propofol infusion was increased to 200 $\mu g \cdot kg^{-1} \cdot min^{-1}$. Clinical signs, such as lack of voluntary movement, stable blood pressure, heart rate and adequate respiration were supportive of an adequate depth of anaesthesia. Since the patient continued to develop dystonic movements two boluses of 5 mg diazepam (Diazemuls®) *iv* were administered with good effect. The propofol infusion was continued at 200 $\mu g \cdot kg^{-1} \cdot min^{-1}$. After a further 30 min the dystonia recurred. Diazepam 10 mg *iv* did not abolish the movements and 100 mg thiopentone *iv* was administered with good effect. A concomitant infusion of thiopentone 1% (in a 50 ml mini-bag) titrated to effect manually was administered for the remaining 20 min of the procedure. A total dose of 500 mg thiopentone and 2448 mg propofol were administered during the period. However, control of dystonia was incomplete and the patient continued to exhibit minor dystonic movements every two to three minutes. The patient did not exhibit any evidence of opisthotonus or nystagmus at any time.

During the first 45 min of the procedure the patient was under deep sedation and, for the remainder, under deep anaesthesia. The patient breathed spontaneously during the MRI without apnoeic episodes and vital signs were within normal limits. The recovery period was uneventful without obvious neurological changes. There were no concerns for this patient postoperatively and he was discharged the same day at which time his torticollis was in its usual form and frequency. The MRI scan did not demonstrate a surgically resectable focus.

Discussion

Torticollis is the most frequent and familiar type of focal dystonia.[1] The occurence of dyskinetic neck muscle movement that causes abnormal head posture is a distinguishing characteristic of this symptom complex. Involuntary activity involves the sternocleidomastoid, trapezius, and scalenus muscles in sustained contractions that result in slow, twisting, turning movements of the head (torticollis) or, less often, forward flexion (antero-collis) or forceful extention (retro-collis). Similar activity may spread to facial and brachial musculature. Some long term observations indicate that about 65% of patients develop additional dystonic features within 10 yr of its onset.[2] The aetiology of dystonia in this patient was focal cerebral trauma. The patient required investigation with an MRI scan to determine if there was a surgically resectable focus. A MEDLINE® search back to 1965 employing the mesh words ANAESTHESIA, TORTICOLLIS and HYPERREFLEXIA failed to reveal any reports relating to the anaesthetic management of torticollis-dystonia disorder. In a textbook of anaesthesia there was one report suggesting that nitrous oxide at analgesic concentrations could improve the dystonic movements.[3] Successful MRI scanning mandates absence of head and neck movements during the procedure. We used propofol because, in our radiology suite, it is the agent of choice for all patients who require sedation or anaesthesia and it has also been used successfully in the treatment of acute torticollis.[4]

Recently, attention has focused on the relationship between propofol and perioperative neurological sequelae ranging from epileptiform movements to frank seizures and syndromes of decerebrate rigidity.[5,6] The origin of these excitatory effects depends on the balance of excitatory and depressive effects of the anaesthetic agent on neurones within the cortex, thalamus and reticular formation. Somatosensory stimulation (tactile, visual or auditory) has been associated with augmentation of these excitatory effects. In three cases, excitatory sequelae induced by propofol underwent episodic augmentation in conjunction with blood pressure measurement.[6] In our patient an automated blood pressure measurement was made every three minutes and this might explain the frequency of dystonic movements during the latter part of the anaesthetic. It has been proposed that propofol produces a strychnine-like antagonism of glycinergic inhibition.[7] Propofol has also been shown to potentiate glycinergic and GABAergic transmission.[5,7] It is speculated that excitatory phenomena during recovery

from propofol may be due to 'rebound' with impaired inhibition at brain and spinal levels. During propofol anaesthesia there may be 'acute tolerance' with refractoriness in glycinergic and GABAergic pathways. The impaired inhibition of spinal output would account for excitatory effects. Our patient had an intermittent response to propofol with initial control of dystonia and subsequent re-emergence of refractory dystonia under deep propofol anaesthesia. Such a response is compatible with the above hypothesis and consistent with the biphasic convulsive responses to propofol in mice.[5]

Torticollis and dystonia during propofol anaesthesia have not been previously reported. Postoperative torticollis and opisthotonus have been described after fentanyl, enflurane and nitrous oxide anaesthesia.[8] In this case there was a partial response to thiopentone, diazepam and diphenhydramine. It has been suggested that small doses of benzodiazepines should be considered as the first line treatment for such excitatory side-effects of propofol.[7] However, these agents were of limited benefit in our patient. The rationale for such a strategy is that benzodiazepines potentiate activity of $GABA_A$ receptors.[7] In addition, the partial success of thiopentone is consistent with previous reports where thiopentone was found to control propofol induced seizures and opisthotonus only briefly.[8] Physostigmine has also been suggested as a therapeutic choice because it may restore spinal inhibition.[7] It is unlikely that nitrous oxide will have a therapeutic benefit in patients with torticollis because it has also been implicated in the development of postoperative torticollis.[9] In addition, human volunteers and mice have been reported to develop opisthotonus and excitatory phenomena after nitrous oxide administration.[10,11] However, nitrous oxide may temporarily ameliorate but not abolish spasmodic torticollis.[12] These inconsistent reports of the effects of nitrous oxide are also seen with other volatile anaesthetics and suggest that these agents may not blunt all supraspinal-mediated muscle activity and, in some situations, may facilitate neurotransmission in some pathways (example: dopamine mediated nigrostriatal-basal ganglia pathway) to result in exacerbation of symptoms.[13]

Considering our experience and the known effects of propofol we suggest that, propofol should be avoided in patients with torticollis and dystonias. Where complete control of movements is required, as in our patient, it may be necessary to consider general endotracheal anaesthesia in conjunction with muscle relaxants.

References

1 *Flint Beal M, Richardson EP, Martin JB.* Degenerative diseases of the nervous system. *In:* Wilson JD, Brunwald E, Isselbacher KJ, *et al.* (Eds.). Principles of Internal Medicine. New York: McGraw Hill Inc, 1991: 2060–9.

2 *Rowland LP.* Merritt's Text book of Neurology, 8th ed. 1989: 655–6.

3 *Martz DG, Schreibman DL, Matjasko MJ.* Neurological diseases. In: Katz J, Benumof JL, Kadis LB (Eds.). Anesthesia and Uncommon Diseases. Philadelphia: W.B.Saunders Co, 1990: 560–86.

4 *Borgeat A.* Usefulness of propofol in torticollis (Letter). Br J Anaesth 1991; 66: 530.

5 *Bevan JC.* Propofol-related convulsions (Editorial). Can J Anaesth 1993; 40: 805–9.

6 *Saunders PR, Harris MN.* Opisthotonus and other unusal neurological sequelae after outpatient anaesthesia. Anaesthesia 1990; 45: 552–7.

7 *Ries CR, Scoates PJ, Puil E.* Opisthotonus following propofol: a nonepileptic perspective and treatment strategy. Can J Anaesth 1994; 41: 414–9.

8 *DeFriez CB, Wong HC.* Seizures and opisthotonus after propofol anesthesia. Anesth Analg 1992; 75: 630–2.

9 *Dehring DJ, Gupta B, Peruzzi WT.* Postoperative opisthotonus and torticollis after fentanyl, enflurane and nitrous oxide. Can J Anaesth 1991; 38: 919–25.

10 *Hornbein TF, Eger EI II, Winter PM, Smith G, Westone D, Smith KH.* The minimum alveolar concentration of nitrous oxide in man. Anesth Analg 1982; 61: 553–6.

11 *Rupreht J, Dworacek B, Ducardus R, Schmitz PI, Dzoljic MR.* The involvement of the central cholinergic and endorphinergic systems in the nitrous oxide withdrawal syndrome in mice. Anesthesiology 1983; 58: 524–6.

12 *Gillman MA, Sandyk R.* Nitrous oxide ameliorates spasmodic torticollis. Eur Neurol 1985; 24:292–3.

13 *Stemp LI, Taswell C.* Spastic torticollis during general anesthesia: case report and review of receptor mechanisms. Anesthesiology 1991; 75: 365–6.