

# Anesthetic Considerations of Sporadic Inclusion Body Myositis in an Elderly Man With Orthopedic Trauma

Dominik T Steck,<sup>1</sup> Christine Choi,<sup>1</sup> Suneeta Gollapudy,<sup>1</sup> and Paul S Pagel<sup>2,\*</sup>

<sup>1</sup>Department of Anesthesiology, Medical College of Wisconsin, Milwaukee, Wisconsin, USA

<sup>2</sup>Anesthesia Service, Clement J Zablocki Veterans Affairs Medical Center, Milwaukee, Wisconsin, USA

\*Corresponding author: Paul S Pagel, Anesthesia Service, Clement J Zablocki Veterans Affairs Medical Center, Milwaukee, Wisconsin, USA. Tel: +414-3842000, Fax: +414-9025479, E-mail: pspagel@mcw.edu

Received 2015 December 19; Accepted 2016 January 26.

## Abstract

**Introduction:** Sporadic inclusion body myositis (IBM) is an inflammatory myopathy characterized by progressive asymmetric extremity weakness, oropharyngeal dysphagia, and the potential for exaggerated sensitivity to neuromuscular blockers and respiratory compromise. The authors describe their management of a patient with IBM undergoing urgent orthopedic surgery.

**Case Presentation:** An 81-year-old man with IBM suffered a left intertrochanteric femoral fracture after falling down stairs. His IBM caused progressive left proximal lower extremity, bilateral distal upper extremity weakness (left > right), and oropharyngeal dysphagia (solid food, pills). He denied dyspnea, exercise intolerance, and a history of aspiration. Because respiratory insufficiency resulting from diaphragmatic dysfunction and prolonged duration of action of neuromuscular blockers may occur in IBM, the authors avoided using a neuromuscular blocker. After applying cricoid pressure, anesthesia was induced using intravenous lidocaine, propofol, remifentanyl followed by manual ventilation with inhaled sevoflurane in oxygen. Endotracheal intubation was accomplished without difficulty; anesthesia was then maintained using remifentanyl and sevoflurane. The fracture was repaired with a trochanteric femoral nail. The patient was extubated without difficulty and made an uneventful recovery.

**Conclusions:** In summary, there is a lack of consensus about the use of neuromuscular blockers in patients with IBM. The authors avoided these drugs and were able to easily secure the patient's airway and maintain adequate muscle relaxation using a balanced sevoflurane-remifentanyl anesthetic. Clinical trials are necessary to define the pharmacology of neuromuscular blockers in patients with IBM and determine whether use of these drugs contributes to postoperative respiratory insufficiency in these vulnerable patients.

**Keywords:** Acquired Muscle Disease, Inclusion Body Myositis, Inflammatory Myopathy, Orthopedic Surgery, Trauma

## 1. Introduction

Sporadic inclusion body myositis (IBM) is a rare acquired idiopathic inflammatory myopathy with autoimmune and neurodegenerative features that is characterized by slowly progressive asymmetric weakness of the proximal lower extremity (quadriceps) or distal upper extremity (wrist and distal phalangeal flexors) without sensory deficits (1, 2). IBM is most commonly observed in men over fifty years of age. The incidence of IBM is approximately 8 per million in the United States (3) and ranges between 2 and 12 per million in other parts of the world (1, 4). The slow onset of the disease often delays its diagnosis for several years (5). Involvement of the quadriceps muscle group contributes to gait instability and falls in the vast majority of patients (6). Oropharyngeal dysphagia also occurs frequently and increases the risk of aspiration in patients with IBM because clearance of secretions may be impaired (7, 8). In contrast to other inflammatory myopathies (e.g., polymyositis, dermatomyositis), intrinsic pulmonary disease (e.g., interstitial pneumonitis, alveolitis) and myocardial fibrosis compli-

cated by arrhythmias or heart failure does not occur in IBM. Nevertheless, diaphragmatic dysfunction may contribute to respiratory insufficiency in some patients with IBM (9).

A clinical history of weakness exceeding twelve months in duration, elevated plasma creatine phosphokinase concentration, and muscle histology demonstrating endomysial inflammation, rimmed vacuoles, and amyloid or other protein deposition are required to establish the diagnosis of IBM (10). Magnetic resonance imaging may also provide confirmatory evidence of the disease (11). The etiology of IBM remains undefined, but autoimmunity against cytosolic 5'-nucleotidase 1A has been implicated in the disease's pathophysiology (12). Coincident autoimmune diseases (e.g., systemic lupus erythematosus, Sjogren's syndrome, sarcoidosis) may also be present. A neurodegenerative cause of IBM has also been proposed for which considerable indirect evidence exists (1). Treatment for IBM is entirely supportive because the disease is essentially unresponsive to the immunosup-

pressive therapies used to manage other inflammatory myopathies (13-15). Few reports exist that comment on the anesthetic considerations of IBM, but concern about the potential for exaggerated sensitivity to neuromuscular blockers and respiratory compromise has been consistently expressed. The authors describe their perioperative management of an elderly man with IBM undergoing urgent surgery after sustaining orthopedic trauma and discuss the anesthetic ramifications of the disease.

## 2. Case Presentation

An 81-year-old, 77 kg, 183 cm man with a three-year history of muscle biopsy-proven IBM suffered a left intertrochanteric femoral fracture and a small parafalcine subdural hematoma as a result of falling down stairs in his home. The patient originally had progressive left proximal lower extremity and bilateral distal upper extremity weakness (left > right) that began several years before the diagnosis was definitively established with a muscle biopsy. The patient's serum creatine phosphokinase concentration was 748 U/L (normal < 170 U/L) at the time of the diagnosis. The patient had several near falls in the months preceding the accident. His left hand and wrist weakness affected his activities of daily living to some degree (fine motor skills), but physical and occupational therapy helped the patient remain active and he continued to live independently with occasional assistance from his wife. Because of worsening left quadriceps weakness, he had recently begun using a cane to help him rise from a seated position. The patient had a history of oropharyngeal dysphagia (solid food, pills) for which he was receiving care from an otolaryngologist. The patient also had chronic gastroesophageal reflux disease that was well controlled with a proton pump inhibitor. He did not have difficulty swallowing liquids and denied respiratory complaints associated with chronic aspiration. The patient did not lose consciousness after striking his head on the stairs and had no new neurological complaints. His other past medical history was notable for coronary artery disease, hyperlipidemia, chronic low back pain, and vertebral artery stenosis. He underwent coronary artery bypass graft surgery 16 years before the current admission. A previous transthoracic echocardiography revealed normal left ventricular systolic function. He denied chest pain, dyspnea, and exercise intolerance. The physical examination and radiographs of his femur revealed the left intertrochanteric fracture. The Glasgow coma scale was 15 and there were no new focal neurological deficits. A magnetic resonance imaging study demonstrated the small parafalcine subdural hematoma without a cerebral mass effect.

The patient was transported to the operating room for repair of the left intertrochanteric fracture. Because primary respiratory failure (16, 17) resulting, at least in part, from diaphragmatic dysfunction (9) has been previously reported in patients with IBM and prolonged duration of

action of neuromuscular blockers may also occur, the authors opted to avoid the use of a neuromuscular blocker for endotracheal intubation and during maintenance of anesthesia in the current patient. After applying cricoid pressure, anesthesia was induced using intravenous lidocaine (0.5 mg.kg<sup>-1</sup>), propofol (1 mg.kg<sup>-1</sup>), and remifentanyl (infusion of 0.1 mcg.kg<sup>-1</sup>.min<sup>-1</sup>). Manual positive-pressure ventilation with sevoflurane (inspired concentrations between 2% and 3%) in oxygen (100%) was used with sustained cricoid pressure to supplement to intravenous anesthetics after loss of consciousness. After uneventful endotracheal intubation, anesthesia was maintained with sevoflurane (end-tidal concentrations between 0.7% and 1.5%) and remifentanyl (infusion rates of 0.05 to 0.15 mcg.kg<sup>-1</sup>.min<sup>-1</sup>). The orthopedic surgeon repaired the fracture with a trochanteric femoral nail. Intravenous acetaminophen and fentanyl were administered for postoperative analgesia. The patient tolerated the procedure well. He demonstrated vigorous spontaneous respiratory effort as he emerged from anesthesia and was extubated without difficulty in the operating room. The patient made an uncomplicated recovery from surgery and was subsequently transferred to a rehabilitation facility in stable condition.

## 3. Discussion

Only a few reports have described the anesthetic implications of IBM, which is somewhat surprising considering that patients with this disease often sustain injuries as a result of falls (6) or require cricopharyngeal procedures (e.g., surgical myotomy, botulinum toxin injection) for treatment of oropharyngeal dysphagia (7, 18, 19). Ntatsaki et al. discussed their anesthetic technique for a patient with IBM who required serial electroconvulsive therapy (ECT) for treatment of severe psychotic depression (20). These authors used a reduced dose of mivacurium to facilitate endotracheal intubation during each ECT because they were concerned about sensitivity to neuromuscular blockers and postoperative respiratory complications. The approach provided satisfactory intubating conditions, but it did modestly delay emergence after ECT because the duration of neuromuscular blockade exceeded the length of the short procedure. The patient suffered no adverse sequelae. In contrast, two reports published in the Japanese literature emphasized that IBM may be associated with postoperative pulmonary complications after general anesthesia. Igari and colleagues used a reduced dose of rocuronium in a patient with IBM undergoing video-assisted thoracic surgery during propofol-remifentanyl anesthesia because they were concerned about postoperative respiratory depression (21). Indeed, the authors were unable to extubate their patient after reversal of neuromuscular blockade, most likely because of underlying pulmonary dysfunction related to IBM. Similar to the authors' approach in the current patient, Nakano et al described endotracheal intubation without

the use of a neuromuscular blocker after induction of general anesthesia in an elderly man with IBM undergoing a jejunostomy, but this patient developed postoperative aspiration pneumonia (22). The current patient did not have a history of compromised pulmonary function and was easily extubated after surgery. His oropharyngeal dysphagia also did not interfere with his ability to swallow liquids nor did he describe a history consistent with chronic aspiration. Indeed, he did not develop aspiration pneumonia after surgery. Marinho et al. reported a unique approach to anesthesia in a patient with IBM who underwent percutaneous thoracic vertebral kyphoplasty in prone position (23). This patient required chronic non-invasive ventilation because of poor respiratory function. The patient's intraoperative care was managed using epidural anesthesia, conscious sedation, and bi-level positive airway pressure. In this case, the authors (23) avoided endotracheal intubation entirely because they were concerned that the patient's preexisting respiratory dysfunction may preclude their ability to wean the patient from mechanical ventilation after the procedure.

The current and previous (20-23) cases used reduced doses of or completely avoided neuromuscular blockers in patients with IBM because of theoretically enhanced sensitivity to these drugs, but with the exception of anecdotal experience, there are no data to definitively establish the pharmacokinetics, pharmacodynamics, and relative safety of neuromuscular blockers in this setting. The actions of neuromuscular blockers may be prolonged in other forms of inflammatory myopathy, but this contention has not been examined systematically in controlled clinical trials and remains controversial, in part because inflammatory myopathies including IBM may not affect the functional integrity of the neuromuscular junction (1). Flusche et al. reported that recovery from vecuronium was significantly delayed in an elderly man with polymyositis (24), but Brown et al. reported that the durations of action of succinylcholine and atracurium both fell within the normal range in a woman with dermatomyositis (25). Johns et al. described an abnormal contracture response to succinylcholine in a 2 year-old boy with dermatomyositis, but the duration of action of the depolarizing neuromuscular blocker was not prolonged (26). A rise in serum potassium concentration was also observed in this case, but the effect was not overly exaggerated (26), suggesting that succinylcholine may not precipitate dangerous hyperkalemia in patients with inflammatory myositis. Nevertheless, the use of succinylcholine is relatively contraindicated in patients with other primary muscle diseases because of an exaggerated hyperkalemic response (27), and for this reason, the authors did not use the depolarizing neuromuscular blocker in the current patient.

In summary, there is a lack of consensus about the use of neuromuscular blockers in patients with IBM and other inflammatory myopathies. The authors avoided these drugs and were able to easily secure the patient's

airway and maintain adequate muscle relaxation using a balanced sevoflurane-remifentanyl anesthetic. The current patient did not have difficulty swallowing liquids or handling secretions despite his oropharyngeal dysphagia nor did he have a history of aspiration, respiratory insufficiency, or diaphragmatic dysfunction. The presence of any of these IBM-related conditions most likely would have prompted the authors to secure the patient's airway using a neuromuscular blocker despite the theoretical risk for a prolonged duration of action. Clinical trials are necessary to define the pharmacology of neuromuscular blockers in patients with IBM and determine whether use of these drugs contributes to postoperative respiratory insufficiency in these vulnerable patients.

## Footnotes

**Authors' Contribution:** Dominik T Steck cared for the patient for the patient in the operating room and wrote drafts of the manuscript. The author has read the final draft of the manuscript and approves it for submission. Christine Choi cared for the patient for the patient in the operating room and wrote drafts of the manuscript. The author has read the final draft of the manuscript and approves it for submission. Suneeta Gollapudy cared for the patient for the patient in the operating room and wrote drafts of the manuscript. The author has read the final draft of the manuscript and approves it for submission. Paul S Pagel wrote and edited drafts of the manuscript. The author has read the final draft of the manuscript and approves it for submission.

**Conflict of Interest:** The authors have no conflicts of interest pursuant to this report.

**Funding/Support:** This work was supported entirely by departmental funds.

## References

1. Dimachkie MM, Barohn RJ. Inclusion body myositis. *Neurol Clin.* 2014;**32**(3):629-46. doi: 10.1016/j.ncl.2014.04.001. [PubMed: 25037082]
2. Lahouti AH, Amato AA, Christopher-Stine L. Inclusion body myositis: update. *Curr Opin Rheumatol.* 2014;**26**(6):690-6. doi: 10.1097/BOR.0000000000000116. [PubMed: 25215417]
3. Wilson FC, Ytterberg SR, St Sauver JL, Reed AM. Epidemiology of sporadic inclusion body myositis and polymyositis in Olmsted County, Minnesota. *J Rheumatol.* 2008;**35**(3):445-7. [PubMed: 18203321]
4. Phillips BA, Zilko PJ, Mastaglia FL. Prevalence of sporadic inclusion body myositis in Western Australia. *Muscle & Nerve.* 2000;**23**(6):970-2. doi: 10.1002/(sici)1097-4598(200006)23:6<970::aid-mus20>3.3.co;2-9. [PubMed: 10842277]
5. Lindberg C, Persson LI, Bjorkander J, Oldfors A. Inclusion body myositis: clinical, morphological, physiological and laboratory findings in 18 cases. *Acta Neurol Scand.* 1994;**89**(2):123-31. [PubMed: 8191875]
6. Hiscock A, Dewar L, Parton M, Machado P, Hanna M, Ramdharry G. Frequency and circumstances of falls in people with inclusion body myositis: a questionnaire survey to explore falls management and physiotherapy provision. *Physiotherapy.* 2014;**100**(1):61-5. doi: 10.1016/j.physio.2013.06.002. [PubMed: 23954023]
7. Darrow DH, Hoffman HT, Barnes GJ, Wiley CA. Management of dysphagia in inclusion body myositis. *Arch Otolaryngol Head Neck*

- Surg.* 1992;**118**(3):313-7. [PubMed: 1313247]
8. Houser SM, Calabrese LH, Strome M. Dysphagia in patients with inclusion body myositis. *Laryngoscope.* 1998;**108**(7):1001-5. [PubMed: 9665246]
  9. Teixeira A, Cherin P, Demoule A, Levy-Soussan M, Straus C, Verin E, et al. Diaphragmatic dysfunction in patients with idiopathic inflammatory myopathies. *Neuromuscul Disord.* 2005;**15**(1):32-9. doi: 10.1016/j.nmd.2004.09.006. [PubMed: 15639118]
  10. Hilton-Jones D, Miller A, Parton M, Holton J, Sewry C, Hanna MG. Inclusion body myositis: MRC Centre for Neuromuscular Diseases, IBM workshop, London, 13 June 2008. *Neuromuscul Disord.* 2010;**20**(2):142-7. doi: 10.1016/j.nmd.2009.11.003. [PubMed: 20074951]
  11. Cox FM, Reijnierse M, van Rijswijk CS, Wintzen AR, Verschuuren JJ, Badrising UA. Magnetic resonance imaging of skeletal muscles in sporadic inclusion body myositis. *Rheumatology (Oxford).* 2011;**50**(6):1153-61. doi: 10.1093/rheumatology/ker001. [PubMed: 21288962]
  12. Benjamin Larman H, Salajegheh M, Nazareno R, Lam T, Sauld J, Steen H, et al. Cytosolic 5'-nucleotidase 1A autoimmunity in sporadic inclusion body myositis. *Annals Neurology.* 2013;**73**(3):408-18. doi: 10.1002/ana.23840.
  13. Barohn RJ, Amato AA, Sahenk Z, Kissel JT, Mendell JR. Inclusion body myositis: explanation for poor response to immunosuppressive therapy. *Neurology.* 1995;**45**(7):1302-4. [PubMed: 7617187]
  14. Muscle Study Group. Randomized pilot trial of INF1a (Avonex) in patients with inclusion body myositis. *Neurology.* 2001;**57**(9):1566-70. doi: 10.1212/wnl.57.9.1566. [PubMed: 11706093]
  15. Barohn RJ, Herbelin L, Kissel JT, King W, McVey AL, Saperstein DS, et al. Pilot trial of etanercept in the treatment of inclusion-body myositis. *Neurology.* 2006;**66**(2 Suppl 1):S123-4. doi: 10.1212/01.wnl.0000192258.32408.54. [PubMed: 16432140]
  16. Cohen R, Lipper S, Dantzker DR. Inclusion body myositis as a cause of respiratory failure. *Chest.* 1993;**104**(3):975-7. [PubMed: 8396004]
  17. Voermans NC, Vaneker M, Hengstman GJD, ter Laak HJ, Zimmerman C, Schelhaas HJ, et al. Primary respiratory failure in inclusion body myositis. *Neurology.* 2004;**63**(11):2191-2. doi: 10.1212/01.wnl.0000145834.17020.86. [PubMed: 15596785]
  18. Danon MJ, Friedman M. Inclusion body myositis associated with progressive dysphagia: treatment with cricopharyngeal myotomy. *Can J Neurol Sci.* 1989;**16**(4):436-8. [PubMed: 2553229]
  19. Liu LW, Tarnopolsky M, Armstrong D. Injection of botulinum toxin A to the upper esophageal sphincter for oropharyngeal dysphagia in two patients with inclusion body myositis. *Can J Gastroenterol.* 2004;**18**(6):397-9. [PubMed: 15190396]
  20. Ntatsaki E, D'Mello O, Lewis J, Underwood BR, Smith M, Head L. Electroconvulsive treatment for a patient with psychotic depression and inclusion body myositis. *J ECT.* 2009;**25**(2):125-8. doi: 10.1097/YCT.0b013e31817e0ef6. [PubMed: 18708945]
  21. Igari Y, Ito Y, Nagaya K. [Anesthesia for pneumothorax surgery in a patient with type II chronic respiratory failure associated with inclusion body myositis]. *Masui.* 2014;**63**(2):172-4. [PubMed: 24601112]
  22. Nakano N, Satsumae T, Mizutani T, Kimura M, Tokuwaka J, Tanaka M. Anesthetic Management for a Patient with Inclusion Body Myositis. *J Clin Anesth.* 2012;**32**(5):809-13. doi: 10.2199/jjcsa.32.809.
  23. Marinho A, Guimarães M, Lages NCR, Correia C. Role of noninvasive ventilation in perioperative patients with neuromuscular disease: a clinical case. *Rev Bras Anesthesiol.* 2014.
  24. Flusche G, Unger-Sargon J, Lambert DH. Prolonged neuromuscular paralysis with vecuronium in a patient with polymyositis. *Anesth Analg.* 1987;**66**(2):188-90. [PubMed: 2880531]
  25. Brown S, Shupak RC, Patel C, Calkins JM. Neuromuscular blockade in a patient with active dermatomyositis. *Anesthesiology.* 1992;**77**(5):1031-3. [PubMed: 1443721]
  26. Johns RA, Finholt DA, Stirt JA. Anaesthetic management of a child with dermatomyositis. *Can Anaesth Soc J.* 1986;**33**(1):71-4. [PubMed: 3948051]
  27. Segura LG, Lorenz JD, Weingarten TN, Scavonetto F, Bojanic K, Selcen D, et al. Anesthesia and Duchenne or Becker muscular dystrophy: review of 117 anesthetic exposures. *Paediatr Anaesth.* 2013;**23**(9):855-64. doi: 10.1111/pan.12248. [PubMed: 23919455]