

Anesthetic Management of a Patient with Hallervorden-Spatz Disease

RAYMOND C. ROY, PH.D., M.D.,* STEPHEN MCLAIN, S.R.N.A.,†
ANNE WISE, C.R.N.A.,‡ LOUIS DES. SHAFFNER, M.D.§

Hallervorden-Spatz disease (HSD) is a rare autosomal recessive disorder of the basal ganglia which follows a slowly progressive course from its onset in late childhood to death about ten years later.^{1,2} Dementia and dystonia with torticollis, scoliosis, and oromandibular rigidity are commonly present.³ There is no specific laboratory test diagnostic for this condition and no effective treatment is known. The diagnosis of HSD can only be made at autopsy unless a similarly affected sibling has died and demonstrated iron pigmentation in the globus pallidus and substantia nigra and widely disseminated focal axonal swelling.^{4,5} Approximately 75 cases of HSD have been reported since its initial description in 1922. There has been no discussion of anesthetic experience.

REPORT OF A CASE

A 20-year-old, 41-kg woman presented for closure of a leaking and excoriated gastrostomy and placement of a Dobhoff feeding tube. An older sister died of HSD.

Her past medical history was significant for athetosis and dystonia of seven years' duration which confined her to bed. Oromandibular rigidity prevented speaking and chewing. The increased tone of the muscles of deglutition made swallowing of food and secretions almost impossible. A feeding gastrostomy created under local anesthesia at age 14 years provided access for home nutritional support. She had been hospitalized on numerous occasions for the treatment of pneumonia. Over the previous two months, 120 mg diazepam, 150 mg dantrolene, 600 mg thiorazine, 80 mg baclofen, and 150 mg diphenhydramine in divided daily doses only moderately attenuated her agitation and dystonia. Nightly choral hydrate ameliorated her posturing enough to permit some passive movement of her head and to unclench her teeth to allow suctioning of her oropharynx.

One week prior to her surgery she presented with a left lower lobe pneumonia: rectal temperature 40° C; respiratory rate 40 breaths·min⁻¹; and leukocyte count 12,500 cells·mm⁻³. With a FI_O₂ of 0.4 by face shield, pH_a was 7.48, PaO₂ 82 mmHg, and PaCO₂ 54 mmHg. But

two months earlier, pH_a was 7.48, PaO₂ 85 mmHg and PaCO₂ 26 mmHg with an oral temperature of 38.3° C and respiratory rate 28 breaths·min⁻¹ while breathing room air. She was treated with intravenous penicillin, chest physiotherapy, and spontaneous nebulization with isoetharine. A 28-French nasopharyngeal airway was inserted to facilitate suctioning.

Physical examination prior to surgery revealed torticollis to the left, severe scoliosis, and such marked oromandibular rigidity that her mouth could not be opened (fig. 1). Her posturing intensified with noxious stimulation. Rectal temperature was 38° C. Respiratory rate was 24 breaths·min⁻¹. Auscultation of her lungs revealed diffuse rhonchi. She could not cough effectively.

She received her regular doses of 25 mg dantrolene, 20 mg diazepam, 100 mg thiorazine, and 25 mg diphenhydramine three hours earlier through her gastrostomy tube. Anesthesia was induced with halothane and nitrous oxide by inhalation through a mask with spontaneous ventilation. The airway was easily maintained. With increasing depth of anesthesia the torticollis, scoliosis, and oromandibular rigidity gradually disappeared. Laryngoscopy with a Miller 2 blade was performed easily 20 minutes later with spontaneous ventilation during deep halothane-oxygen anesthesia. The trachea was intubated with a 24-French, high-volume, low-pressure, cuffed endotracheal tube and ventilation was controlled. The inspired gases were humidified. Adequate muscle relaxation was obtained from halothane. No neuromuscular blockers were administered. There was no cardiovascular instability. Intraoperative monitoring included a blood pressure cuff, precordial stethoscope, electrocardiogram, and rectal temperature probe.

At the conclusion of the surgical procedure the patient was transported to the recovery room breathing spontaneously through a T-bar with the endotracheal tube still in place. On emergence, the dystonic posturing returned. Her trachea was extubated two hours postoperatively in the recovery room. With a FI_O₂ of 0.4 by face shield, pH_a was 7.43, PaO₂ 150 mmHg and PaCO₂ 42 mmHg with a rectal temperature of 37.6° C and a respiratory rate of 28 breaths·min⁻¹. A 28-French nasopharyngeal airway was reinserted and her regular medications resumed in the recovery room through the feeding tube. The patient returned home on postoperative day seven without sequelae.

DISCUSSION

Extensive experience with basal ganglia disorders, in general,^{6,7} and dystonia musculorum deformans (torsion dystonia), in particular,^{7,8} suggests that extrapyramidal signs, such as chorea, athetosis, dystonia, rigidity, and tremor, disappear with the induction of anesthesia and reappear on emergence. Amelioration of this patient's posturing with chloral hydrate suggested that the same also would occur in HSD. But in any patient with torticollis, oromandibular rigidity and scoliosis of long duration, bony or joint changes or muscle contractures may fix the jaw or cervical spine so that mobility could

* Assistant Professor, Department of Anesthesia.

† Student, Nurse Anesthesia.

‡ Instructor, Nurse Anesthesia.

§ Professor, Department of Surgery.

Received from the Departments of Anesthesia and Surgery, North Carolina Baptist Hospital, and Bowman Gray School of Medicine of Wake Forest University, Winston-Salem, North Carolina, 27103. Accepted for publication October 4, 1982.

Address reprint requests to Dr. Roy.

Key words: Muscle, skeletal; dystonia; contractility; Hallervorden-Spatz disease. Brain: basal ganglia; Hallervorden-Spatz disease. Airway: obstruction. Intubation, endotracheal: complications; technique.

be limited even in the presence of deep anesthesia or muscle relaxants.⁷

A standard induction-intubation sequence with thiopental and succinylcholine was avoided in this patient for fear of being unable to maintain a patent airway and of being unable to position her for rapid intubation of her trachea should airway obstruction develop. Awake intubation techniques were not attempted because noxious stimulation intensified her dystonia despite diazepam, dantrolene, thorazine, diphenhydramine, and baclofen. A spontaneous breathing technique was chosen because it provided a way of gradually inducing anesthesia without dramatically altering airway dynamics or respirations with drugs which would take minutes to wear off should the airway be lost. The surgeon was prepared for a tracheostomy. Fortunately, her torticollis and oromandibular rigidity and some of her scoliosis disappeared with anesthesia so that intubation was performed easily.

Halothane alone provided adequate muscle relaxation for endotracheal intubation and surgery. If additional relaxation had been necessary, any of the non-depolarizing agents could have been given.⁶⁻⁸

Although depolarizing muscle relaxants have been administered to patients with torsion dystonia without complication,⁷ succinylcholine is relatively contraindicated in patients with HSD, even though this patient does not appear to fit into any of the five categories of increased skeletal muscle sensitivity described by Gronert and Theye.⁹ There was no evidence for an upper motor neuron lesion, such as increased stretch reflexes or a Babinski response. Despite being bed-ridden, she showed no evidence of muscle wasting; she had been gaining weight because of good nutritional support and she was in a state of constant muscle activity. However, autopsies on other patients with HSD frequently have revealed muscle wasting secondary to poor nutrition and diffuse axonal changes in the brain which may involve upper motor neurons to an unpredictable extent.^{2,5} Therefore, hyperkalemic cardiac arrest with succinylcholine is a possibility. One case of succinylcholine-induced hyperkalemia has been described in a patient with Parkinson's disease.¹⁰

Other variant responses to succinylcholine such as contractures or malignant hyperthermia have not been reported in patients with basal ganglia disorders. The administration of $4 \text{ mg} \cdot \text{kg}^{-1} \cdot \text{day}^{-1}$ dantrolene to this patient complicates any discussion of the risk of malignant hyperthermia in HSD.

Preoperative pulmonary assessment in pediatric neuromuscular disease recently has been reviewed.¹¹ Severe mental retardation and advanced disease in this patient limited assessment to auscultation, respiratory rate, minute ventilation, chest roentgenogram, and arterial blood



FIG. 1. Patient with Hallervorden-Spatz disease demonstrating dystonic posturing with scoliosis and torticollis.

gases. She could not recruit from her inspiratory or expiratory reserve volumes to cough effectively or to breathe more deeply. Her only response to an increased ventilatory requirement was to increase her respiratory rate. Thus, important extubation criteria were a normal PaCO_2 at an acceptable respiratory rate and lungs clear to auscultation at her preoperative level of dystonia. The trachea was extubated two hours after anesthesia was discontinued in order to avoid the loss of airway which might occur during emergence. Prolonged postoperative endotracheal intubation also enhanced effective pulmonary toilet and avoided the respiratory depressant effects of residual halothane including decreased ventilatory responses to hypoxia and acidemia.¹²

In summary, a patient with HSD was anesthetized safely with halothane and her trachea intubated. Her dystonic posturing disappeared upon induction of anesthesia and returned on emergence as with other basal ganglia disorders.

The authors thank Mr. Bruce Coats for photographic assistance.

REFERENCES

1. Dooling EC, Richardson EP, Davis KR: Computed tomography in Hallervorden-Spatz disease. *Neurology* 30:1128-1130, 1980

2. Elejalde BR, de Elejalde MMJ, Lopez FP: Hallervorden-Spatz disease. *Clinical Genetics* 16:1-18, 1979
3. Lakke JPF: Classification of extrapyramidal disorders. *J Neurol Sci* 51:311-327, 1981
4. Goldberg W, Allen N: Nonspecific accumulation of metals in the globus pallidus in Hallervorden-Spatz disease. *Trans Am Neurol Assoc* 104:106-108, 1979
5. Kalyanasundaram S, Srinivas HV, Deshpande DH: An adult with Hallervorden-Spatz disease: clinical and pathological study. *Clin Neurology Neurosurg* 82:245-249, 1980
6. Kadis LB: *Neurologic disorders, Anesthesia and Uncommon Diseases*. Edited by Katz J, Benumof J, Kadis LB. Philadelphia, WB Saunders, 1981, pp 485-489
7. Steen SN: Anesthetic management for basal ganglia surgery in patients with movement disorders. *Anesth Analg (Cleve)* 44:66-69, 1965
8. Davis NL, Davis R: Anesthetic management of a patient with dystonia musculorum deformans. *ANESTHESIOLOGY* 42:630-631, 1975
9. Gronert GA, Theye RA: Pathophysiology of hyperkalemia induced by succinylcholine. *ANESTHESIOLOGY* 43:89-99, 1975
10. Gravlee GP: Succinylcholine-induced hyperkalemia in a patient with Parkinson's disease. *Anesth Analg (Cleve)* 59:444-446, 1980
11. Silva WJ, Rucker RW, Ewald JA: Preoperative pulmonary assessment in pediatric neuromuscular disease. *Anesthesiology Review* 9:35-42, 1982
12. Craig DB: Postoperative recovery of pulmonary function. *Anesth Analg (Cleve)* 60:46:52, 1981

Anesthesiology
58:384-386, 1983

Prolonged Effect of Succinylcholine after Neostigmine and Pyridostigmine Administration in Patients with Renal Failure

MICHAEL J. BISHOP, M.D.,* AND THOMAS F. HORNBEIN, M.D.†

We report two cases of prolonged succinylcholine neuromuscular blockade in patients with renal failure, each of which occurred when the drug was administered several hours after the end of a kidney transplantation. We feel these prolonged blockades can be explained by altered anticholinesterase kinetics, although lower than normal pseudocholinesterase levels also have contributed. That prolonged blockade has not been reported previously in this circumstance is explained by the rare occurrence of a combination of events; the patient with renal failure who has received an anticholinesterase and is administered succinylcholine several hours later. Based on our experience and review of the pertinent literature, when these three conditions are present, pro-

longed succinylcholine blockade probably is the rule rather than the exception.

REPORTS OF TWO CASES

Case 1

A 30-year-old, 70-kg woman with chronic renal failure was anesthetized with halothane and nitrous oxide for a kidney transplantation. The patient had been undergoing peritoneal dialysis three times a week. Intravenous medications administered during the five-hour transplant procedure included 300 mg thiopental, 21 mg *d*-tubocurarine, 40 mg furosemide, 40 mg methylprednisolone, 12.5 g mannitol, and 50 mg diphenhydramine. At the end of the procedure, 1 mg atropine and 2.5 mg neostigmine, iv, successfully antagonized the *d*-tubocurarine neuromuscular blockade. The trachea was extubated and the patient then was taken to the recovery room in satisfactory condition, although she remained anuric.

Two hours after the end of surgery, the patient returned to the operating room because of continued bleeding at the surgical site and persistent anuria. Halothane and nitrous oxide anesthesia were again used, but this time endotracheal intubation was facilitated by the iv administration of 100 mg succinylcholine. One hour later, at the end of the procedure, tetanic stimulation produced marked fade and a phase II neuromuscular block was diagnosed. Ventilation was controlled in the recovery room until full muscular strength returned two hours later.

Serum pseudocholinesterase activity, measured immediately postoperatively was 3.1 IU (normal, 4.3-10.9), and dibucaine and fluoride numbers were both within the normal range.

* Assistant Professor of Anesthesia and Medicine.

† Professor, Departments of Anesthesiology and Physiology and Biophysics; Chairman, Department of Anesthesiology.

Case 2 occurred at the University of California, San Francisco during Dr. Bishop's residency training.

Received from the Departments of Anesthesiology and Medicine, University of Washington, Seattle, Washington.

Address reprint requests to Dr. Bishop: Department of Anesthesiology, ZA-14, Harborview Medical Center, 325 Ninth Avenue, Seattle, Washington 98104.

Key words: Neuromuscular relaxants; succinylcholine. Antagonists, neuromuscular relaxants; neostigmine; pyridostigmine. Kidney: failure; transplantation. Enzymes: pseudocholinesterase.