Anesthetic Management of a Patient with the Shy-Drager Syndrome

CHARLES A. COHEN, M.D.*

In 1960 Shy and Drager described two patients with orthostatic hypotension and primary degenerative disease of the nervous system. The major clinical manifestations of this entity, now known as the Shy-Drager syndrome, are: 1) orthostatic hypotension; 2) parkinsonism; 3) urinary and bowel dysfunction; 4) impaired potency and libido; 5) decreased sweating. About 100 cases have been reported since 1960. A review of these reveals that at least eight patients have undergone surgical operations. Since no analysis of the course of such individuals during anesthesia has been made so far as can be determined, the following case history is presented.

* Resident in Anesthesia, Department of Anesthesiology, University of Pennsylvania School of Medicine, Philadelphia, Pennsylvania 19104.

Supported (in part) by USPHS Grant 5-P01-GM-15430-04 and by USPHS Research Training Grant 5-T01-GM-00215-13, from the National Institutes of General Medical Sciences, National Institutes of Health.

Report of a Case

The patient, a 50-year-old man, had first experienced dizziness and loss of consciousness following prolonged standing in 1964. By 1967 he had noted slowed speech, occasional urinary incontinence, and impotence. In July 1969, because of increasing syncopal episodes, he had to stop working. A diagnosis of multiple sclerosis and diabetes mellitus was tentatively made because of an elevated CSF protein and an abnormal glucose tolerance test.

In early 1970 the patient was admitted to the Hospital of the University of Pennsylvania (HUP). At this time he had severe orthostatic hypotension, parkinsonism, urinary incontinence, an elevated CSF protein with normal sugar and no cells, an abnormal glucose tolerance test, and Battey bacilli (Type III) on sputum culture. He was treated with fludrocortisone (Florinef), hydroxyamphetamine (Paretrine), NaCl, furosemide, and tolbutamide (Orinase). The following values were normal: PBI, serum electrolytes, Ca, P, liver function tests, 24-hour urine collections for VMA and catecholamines, plasma cortisol, nerve conduction time, EEG, EMG, EKG, brain scan, and diffusing capacity of the lung. Vital ca-
pacity was decreased to 3,400 ml (predicted 4,820 ml), and the roentgenogram of the chest showed mild interstitial fibrosis and normal heart size.

Cardiac catheterization was performed, with the following significant results: 1) Valsalva maneuver—failure of blood pressure to rise above the baseline following release of increased airway pressure; 2) cold pressor test—normal increase in peripheral vascular resistance with slight decrease in cardiac output; 3) isoproterenol infusion (4.9 μg/min)—striking rise in blood pressure after initial decrease; 4) exercise in supine position—pulse rate increase from 100 to 108 beats/min, cardiac output increase from 6.2 to 7.9 l/min, systemic vascular resistance decrease, and no blood pressure change; 5) l-norepinephrine infusion—a greater increase in blood pressure than expected for the dose used (7.06 μg/min); 6) carotid sinus pressure—slight slowing elicited by both right and left carotid sinus massage; 7) saline loading—an increase in left ventricular end-diastolic pressure, increased amplitude of the A wave on the left ventricular pressure tracing, some increase in heart rate, unchanged peripheral vascular resistance and blood pressure.

Over the next few months the patient's condition slowly deteriorated. Trimepyrinidyl (Atrane) was started, and later benztrapine (Cogentin) was added. After four months benztrapine was stopped and levodopa was substituted, but it was soon discontinued because of worsening of the postural hypotension despite a very small dose of the drug. The patient's daily activities were severely limited by symptoms resulting from an atomic neurogenic bladder, and he was admitted to HUP on October 6 for a bilateral ureterocleostomy. At this time hemoglobin was 16.5 gm/100 ml, EKG was normal except for a flat T wave in AVL, and blood gases were normal at rest. For a week in the hospital the majority of blood pressure readings were around 120/80 mm Hg (range 180/120 to 100/65).

The patient was brought to the operating room without premedication; preoperative supine blood pressure was 200/120 mm Hg, with a pulse rate of 70 beats/min and a cardiac output of 8.0 l/min. Satisfactory anesthesia failed to develop after an epidural catheter was injected with 375 mg of 1.5 per cent meperidine (Carbocaine). General anesthesia was then induced with 150 mg of thiopental (Pentothal), and the trachea was intubated following 80 mg of succinylcholine (Anectine). Anesthesia was maintained with N2O, O2, and methoxyflurane (using an Ohio "S" vaporizer). In the first 15 minutes the blood pressure declined slowly to 80/60 mm Hg, with a pulse rate of 80 beats/min. The methoxyflurane (MOF) concentration was decreased; the pressure rose to 90/60 mm Hg, but the degree of anesthesia was now inadequate for operation. After 12-14 minutes

of a dilute phenylephrine (Neo-Synephrine) drip intravenously (5 mg per liter of Normosol-R1), the pressure remained unchanged, although cardiac output at this time was 8.4 l/min. The phenylephrine concentration was increased to 10 mg per liter, and arterial pressure rose to 130/80 mm Hg with pulse rate remaining essentially unchanged. The pressor infusion was used for the remainder of the four-hour surgical procedure; it was discontinued 30 minutes before the end of the operation, but blood pressure decreased from 120/80 to 90/60 mm Hg over a 10-minute interval and the infusion was restarted. Total blood loss was 250 ml, and the patient received 1,400 ml of Normosol-R by vein. The phenylephrine infusion was tapered off to zero over the next 90 minutes. The patient was transferred to the surgical intensive care unit with a blood pressure of 108/70 mm Hg and a pulse rate of 92 beats/min. No further pressor infusions were required, and the systolic pressure over the next 24 hours was about 95-100 mm Hg. Intravenous mineralocorticoids and amphetamine were used postoperatively until the patient could be returned to his preoperative medication. He was discharged on October 30.

DISCUSSION

This patient satisfied the criteria for a diagnosis of the Shy-Drager syndrome (SDs). Despite an elevated oral glucose tolerance test, he had a normal nerve conduction time and an absence of sensory findings suggestive of diabetic neuropathy. His clinical course was also unlike that of a diabetic patient with postural hypotension.4

Diffuse involvement of the central and peripheral autonomic systems, the corticobulbar and corticospinal tracts, basal ganglia, and cerebellum has been found in the seven cases of SDs that have come to autopsy.5 Hyper-sensitivity to intravenous norepinephrine has been ascribed to denervation,5 i.e., a denervated structure's being hypersensitive to its physiologic neurotransmitter. However, the observed hypersensitivity to angiotensin5 in some patients with SDs cannot be easily explained by this mechanism, since angiotensin is not known to function as a neurotransmitter. This has led some investigators to postulate defective baroreceptor regulation of blood pressure rather than denervation hypersensitivity as the cause of the postural hypotension.6 The barostatic reflexes in this patient had been shown to be only partially functional, i.e., the heart slowed when the norepi-

† Determined by the dye-dilution technique (indocyanine green) using a Waters cuvette densitometer—calculated by the Stewart-Hamilton technique.

† Abbott Laboratories.
nephrine infusion caused a rise in blood pressure, but there was no pulse increase during postural hypotension and no overshoot following the Valsalva maneuver. As a result of dysfunction of the autonomic nervous system, the clinical evaluation during anesthesia of a patient with SDs is made difficult, because sweating, tachycardia, and blood pressure changes cannot be used reliably as indicators of the depth of anesthesia.

Epidural anesthesia was selected because of the assumption that a partially "sympathectomized" person would have less hypotension with this technique than would a normal individual. There was no evidence of sympathetic blockade or a sensory level following the two injections of mepivacaine that would explain the hypotension observed following induction of general anesthesia. Systemic absorption of 375 mg of mepivacaine appears unlikely to be the cause of the observed hypotension. It is interesting that despite hypersensitivity to norepinephrine preoperatively, the patient responded in a normal manner to phenylephrine. The marked hypotension following light general anesthesia with MOF was due to a decrease in peripheral vascular tone, since the cardiac output did not decrease. This exaggerated hypotensive response to MOF appears to resemble that observed in cats whose baroreflexes have been abolished following section of buffer nerves.2 Halothane does not cause a similar depression of the baroreflex mechanism in identical animal preparations.2 It would seem reasonable, therefore, to expect halothane not to cause unusual hypotension in patients with SDs. One may further speculate that cyclopropane and ether, both of which depress the baroreceptor reflexes in rabbits more than does halothane, may cause marked hypotension in these patients, as did MOF in the present case.

The author thanks Dr. James Hinkle for measuring the cardiac output and Drs. Robert D. Dripps, Donald L. Clark, and Henry L. Price for their advice in the preparation of the manuscript.

REFERENCES


Obstetrics

CARDIOVASCULAR DYNAMICS Serial hemodynamic measurements were made in 17 healthy pregnant women undergoing repeat cesarean section at term under thiopental, nitrous oxide, and succinylcholine anesthesia. Peak cardiac output reached 7 l/min (41 per cent above control values) ten minutes after delivery. A maximum arterial pressure of 131/82 mm Hg (18 per cent above control values) was found just prior to delivery. Peripheral resistance showed little change except for a slight decline past partum. The hemodynamic fluctuations were found to be significantly smaller than those previously reported during cesarean section under subarachnoid block anesthesia and during labor and vaginal delivery under local and caudal anesthesia. From the hemodynamic data presently available, cesarean section under balanced anesthesia should be considered as an alternate method for delivering the infant of the seriously ill, pregnant cardiac patient (Classes III and IV).