

These elevated pressures then stimulate atrial A vagal receptors, which are known to cause peripheral vasodilation.<sup>6</sup> Although this proposal appears sound, no direct evidence for it exists and not all authors agree with this proposed mechanism.

As noted in our case report, the definitive therapy is A-V sequential pacing, which eliminates inappropriate timing of atrial contraction. In situations in which sequential pacing is not available, therapy aimed at restoring a sinus rhythm is indicated. Because the pathophysiologic mechanism of pacemaker syndrome is an inappropriately low systemic vascular resistance, the use of vasopressors and volume expansion in the acute situation would have a strong theoretical basis, provided a sinus rhythm could not be restored.

We have presented a patient with pacemaker syndrome. These patients develop hypotension with the onset of ventricular pacing and may have symptoms of hypotension, including syncope. Although cardiac output falls, it does not fall any more than in control patients. These

patients have normal intravascular volumes and normal cardiovascular responses to the Valsalva maneuver. Reflex vasodilation arising from the stimulation of atrial stretch receptors therefore appears to be responsible for the hypotension. A-V sequential demand pacing is the definitive therapy for this syndrome.

#### REFERENCES

1. Alicandri C, Fouad FM, Tarazi RC, Castle L, Morant V: Three cases of hypotension and syncope with ventricular pacing: Possible role of atrial reflexes. *Am J Cardiol* 42:137-142, 1978
2. Erbel R: Pacemaker syndrome. *Am J Cardiol* 44:771, 1979
3. Lewis ME, Sung RJ, Alter BR, Myerburg RJ: Pacemaker-induced hypotension. *Chest* 79:354-355, 1981
4. Miller M, Fox Stewart, Jenkins R, Schwartz J, Toonder FG: Pacemaker Syndrome: A non-invasive means to its diagnosis and treatment. *Pace* 4:503-506, 1981
5. Samet P, Castillo C, Bernstein WH: Hemodynamic sequelae of atrial, ventricular, and sequential atrioventricular pacing in cardiac patients. *Am Heart J* 72:725-729, 1968
6. Guyton AC: *Textbook of Medical Physiology*. Philadelphia, WB Saunders, 1976, pp 271, 474

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### Precautions in the Anesthetic Management of a Patient with Creutzfeld-Jacob Disease

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Creutzfeld-Jacob Disease (CJD) (subacute spongiform encephalopathy) is a rare, noninflammatory disease of the central nervous system.<sup>1</sup> We describe our anesthetic experience in a patient with CJD and discuss precautions in managing patients with this diagnosis and precautions in handling the equipment used to anesthetize them.

#### REPORT OF A CASE

A 56-year-old man was referred to our hospital for neurologic evaluation. He had been healthy until 8 months ago, at which time he began to experience mild vertigo. In the next 6 months, he subsequently began to have bouts of mild dysphagia and horizontal diplopia. He progressively became more confused and occasionally displayed unusual outbursts of emotion, delusional thinking, and frank hallucinations.

At the time of admission he was suffering from dementia, ataxia, and diffuse myoclonic jerks.

Laboratory values for his blood, urine, and cerebrospinal fluid (CSF) were normal. A computerized axial tomographic scan of the brain revealed only minimal atrophy of the cerebral cortex. An electroencephalogram displayed frequent generalized irregular slowing. He was scheduled for a stereotaxic brain biopsy.

No preoperative medication was given. In addition to disposable masks and hats, both operating room and anesthesia personnel wore gowns and gloves. A 16-gauge catheter was inserted iv in the right arm, and anesthesia was induced with thiopental iv. The trachea was intubated during paralysis induced by pancuronium and anesthesia maintained with 50% oxygen and nitrous oxide. The operation lasted approximately 1 h. Vital signs were stable without autonomic dysfunction. Anesthesia and recovery were uneventful.

All disposable equipment was incinerated and discarded. The laryngoscope then was exposed for 1 h to a 5% solution of sodium hypochlorite. Surgical instruments were autoclaved for 1 h at 121° C (15 PSI). The operating room table, floor, anesthesia machine, and other large surfaces were wiped with the sodium hypochlorite solution.

The brain biopsy revealed neuronal degeneration, gliosis, and vacuolation characteristics of CJD. At the time of this report, the patient has continued to deteriorate.

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#### DISCUSSION

We presented the above case for two reasons. First, there is no documentation of anesthetic experience in a

patient with CJD.<sup>2</sup> Secondly, anesthesiologists should be aware of recommended precautions in the handling of patients with CJD because it is potentially iatrogenically transmissible.

CJD is an extremely rare disease, which is caused by what is believed to be an unconventional transmissible virus. It affects one to two persons per million population and causes approximately 200 deaths per year in the United States. Most cases occur sporadically, but 10% have a family history of presenile dementia.<sup>3</sup> Cathala *et al.*<sup>4</sup> reported one family in which the disease appeared to be transmitted over three generations in an autosomal dominant pattern. Thus, there may be a genetic determination of the expression of the disease. The natural mode of transmission is unknown.

Iatrogenic transmission of CJD has been reported. Duffy *et al.*<sup>5</sup> reported a case in which a patient developed CJD 2 years after receiving a corneal transplant from someone with CJD. Bernoulli *et al.*<sup>6</sup> reported two cases of patients who developed the disease 2.5 and 2.3 years after stereotactic electroencephalographic exploration with silver electrodes previously implanted in a patient with proven CJD. The disease occurred despite the fact that the electrodes had been sterilized with 70% alcohol and formaldehyde.<sup>6</sup> The disease also has been reported in a neurosurgeon, a nurse, three dentists, and three general practitioners.<sup>3</sup> However, no direct link to patients ever has been reported.

The virus has been found not only in the brain and spinal cord of these patients, but also in the lymph nodes, liver, kidney, spleen, lung, cornea, and CSF. Experimentally it has been transmitted to monkeys via intracerebral, subcutaneous, intraperitoneal, intramuscular, and intravenous routes.<sup>7</sup>

Laboratory values are not helpful in the diagnosis of CJD. With the exception of occasional liver dysfunction, there are no abnormalities in the blood or CSF.<sup>8</sup> At first, EEG shows either nonspecific or no changes. Later, there may be high voltage polyphasic discharges that may become periodic at a rate of one or two per second.<sup>9,10</sup>

In contrast to the EEG findings, the somatosensory evoked potentials were found to be normal in two reported cases of CJD. However, more work in this area might be indicated.<sup>3</sup>

There is no known treatment for the disease. Amantadine hydrochloride was reported as a successful form of therapy in two patients.<sup>7</sup> Unfortunately, neither of these patients had pathologic confirmation of the disease. Usually, amantadine is not effective.<sup>7</sup>

When anesthetizing these patients, the diagnosis of CJD should be considered in all patients who have rapid intellectual deterioration and myoclonus, especially when no space-occupying lesions in the brain can be demonstrated. All the patients' tissues should be regarded as

potentially infective, including blood and CSF.<sup>3</sup> If one comes into contact with blood, CSF, nasopharyngeal secretions, urine, or feces, thorough washing of the skin is indicated with a detergent. Vigorous scrubbing with a brush probably should be avoided, as this may abrade the skin. This contact may be avoided by the use of disposable gloves and gowns.<sup>6</sup>

If one accidentally has percutaneous exposure to the tissue of a CJD patient, the wound or puncture should be cleaned thoroughly. Recommended agents are iodine (Betadine®; Purdue Frederick, Norwalk, CT), a phenolic (hexachlorophane or 5% lysol) antiseptic, 0.5% sodium hypochlorite, or a 1:3000 solution of potassium permanganate.<sup>3</sup>

Whenever possible, disposable anesthetic equipment should be used. For nondisposable items, autoclaving for 1 h at temperatures of at least 121° C (15 PSI) remain the method of choice for sterilization. When autoclaving is not possible, a 0.5% solution of sodium hypochlorite may be used.<sup>11</sup> Despite the person-to-person transmissibility of the disease, this risk must be very small by any method other than tissue penetration with contaminated tissues.<sup>11</sup>

In addition, two other findings are important to the anesthesiologist. First, as mentioned above, the patient may have abnormal liver function tests. Second, they may exhibit autonomic dysfunction and may not increase their heart rate in response to atropine. Indeed, postmortem results have indicated alterations in the vagus nerve and sympathetic ganglia patients with CJD.<sup>12</sup>

In conclusion, the anesthesiologist should be aware of the nature and anesthetic needs of the patient with CJD. This knowledge is necessary not only to provide optimal care for the patient, but also to minimize the risk of transmission to himself, his colleagues, and subsequent patients.

#### REFERENCES

1. Shibayama Y, Sakasuchi Y, Nakata K, Toshiyuki G, Nakai M, Takai T, Shirakata S: Creutzfeld-Jacob disease with demonstration of virus-like particles. *Acta Pathol Jpn* 32:695-702, 1982
2. Katz J, Benumof J, Kadis LB: Anesthesia and uncommon diseases. Philadelphia, WB Saunders, 1981, p 489
3. Gajdusek MD, Gibbs CJ, Asher DM, Brown P, Diwan A: Precautions in medical care of, and in handling materials from, patients with transmissible virus dementia (Creutzfeld-Jacob Disease) *N Engl J Med* 297:1253-1258, 1977
4. Cathala F, Chatelain J, Brown P, Dumas M, Gajdusek: Familial Creutzfeld-Jacob Disease. *J Neurol Sci* 47:343-351, 1980
5. Duffy P, Wolf J, Collins G: Possible person-to-person transmission of Creutzfeld-Jacob Disease. *N Engl J Med* 290:692-693, 1974
6. Bernoulli C, Siegfried J, Baumgartner G: Danger of accidental person-to-person transmission of Creutzfeld-Jacob Disease by surgery. *Lancet* 1:478-479, 1977

7. Scully RE, Galdabine JJ, McNeely BU: Case records of the Massachusetts General Hospital, Case 45—1980. *N Engl J Med* 303:1162-1171, 1980
8. Monreal J, Collins GH, Masters CL, Fisher CM, Kin RC, Gibbs CJ, Gajdusek DC: Creutzfeldt-Jacob Disease in an adolescent. *J Neurol Sci* 52:341-350, 1981
9. Behan PO: Creutzfeldt-Jacob Disease. *Br Med J* 284:1658-1659, 1982
10. Traub RD, Pedley TA: Virus-induced electronic coupling: hypothesis on the mechanism of periodic EEG discharges in Creutzfeldt-Jacob disease. *Ann Neurol* 10:405-408, 1981
11. Brown P, Gibbs CJ, Amyx HL, Kingsbury DT, Rohwer RG, Sulima MP, Gajdusek DC: Chemical disinfection of Creutzfeldt-Jacob disease virus. *N Engl J Med* 306:1279-1281, 1982
12. Khurana RK, Garcia JH: Autonomic dysfunction in subacute spongiform encephalopathy. *Arch Neurol* 38:114-117, 1981

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## Unusual Cause of Hypotension after Coronary Artery Bypass Grafting: Idiopathic Hypertrophic Subaortic Stenosis

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Hypotension, decreased cardiac output, and elevated left atrial pressure after discontinuation of extracorporeal circulation for coronary artery bypass grafting (CABG) are usually signs of cardiac failure, which requires treatment with drugs and/or intraaortic counter-pulsation balloon. We recently encountered a case of persistent hypotension worsened by iv catecholamine therapy. A myomectomy was performed to correct hypertrophic subaortic stenosis. The patient then was weaned successfully from cardiopulmonary bypass (CPB).

### REPORT OF A CASE

A 59-year-old 90-kg man, ASA physical status III, was scheduled for elective coronary artery saphenous vein bypass grafting. NYHA class II symptoms of ischemic heart disease and a recent subendocardial myocardial infarction prompted a cardiac catheterization, which revealed three-vessel coronary artery disease (left anterior descending 90%, circumflex 90%, and right coronary artery 50%). The left ventricular end-diastolic pressure was 8 mmHg preangiographic and postangiographic dye injection. The systolic left ventricular and aortic pressures were 170 mmHg. The ventricle was "hyperdynamic," with an ejection fraction of 50%. The mitral valve was normal.

He had a long history of hypertension and hyperuricemia. His physical examination included an enlarged left ventricle and a grade 2/6 systolic ejection murmur, loudest at the second right intercostal space, that radiated to the carotid arteries, bilaterally. The murmur was considered innocent, and the left ventricular hypertrophy secondary to hypertension. Current medications were nitroglycerin 1/150 prn, cutaneous nitroglycerin, hydrochlorothiazide, probenecid, and KCl. ECG showed borderline left ventricular hypertrophy with anterior lateral

ST-T-wave changes compatible with myocardial ischemia. The chest roentgenogram was normal.

About 90 minutes after the administration of diazepam 15 mg po, morphine sulfate 10 mg im, and scopolamine 0.3 mg im, anesthesia was induced with diazepam 0.4 mg/kg iv, lidocaine hydrochloride 1 mg/kg iv, and pancuronium bromide 0.1 mg/kg iv. Nitrous oxide and oxygen were administered via mask before the trachea was intubated. Anesthesia was maintained with fentanyl 20 mcg/kg iv and inhalation of 50% N<sub>2</sub>O and enflurane, 0.7% inspired concentration. Heart rate and rhythm were monitored from V<sub>5</sub> or lead II of the ECG. The left radial and right atrial pressures were monitored continuously before CPB. Also, a 2 F thermister-tipped catheter was inserted into the pulmonary artery via the right ventricle before CPB for determination of cardiac output using the thermodilution technique.

An initial cardiac index measured in duplicate was 2.8 l/min/m<sup>2</sup>. CPB was established, the proximal vein graft anastomoses were made, and the patient was cooled to 28°C. The aorta was cross-clamped and myocardial protection accomplished with potassium cold cardioplegia via the aortic root. There was no unusual amount of regurgitation of cardioplegic solution through the aortic valve during cardioplegia infusion. Seventy-four minutes were required for seven distal vein to coronary artery anastomoses. Total body rewarming was begun during the last distal anastomosis, and once complete, the aortic cross-clamp was removed.

Attempts at weaning the patient from CPB were unsuccessful. Pharmacologic support consisted of nitroglycerin 0.3 mcg · kg<sup>-1</sup> · min<sup>-1</sup> iv and dopamine 10 mcg · kg<sup>-1</sup> · min<sup>-1</sup> iv. Despite this therapy, the mean LAP remained elevated (28 mmHg), and systemic pressure and cardiac output were low, as attempts were made to terminate CPB. Central arterial aortic pressure (100 mmHg) was measured and equaled the radial artery pressure, but a needle placed in the left ventricle revealed a maximum systolic pressure of 180 mmHg (fig. 1A). Premature ventricular contractions were produced by manual stimulation of the heart surface, which showed a postextrasystolic decrease in radial arterial pressure, instead of the usual increase in arterial blood pressure (positive Brockenbrough's sign—fig. 2).<sup>1</sup> A presumptive diagnosis of left ventricular outflow obstruction was made possibly secondary to septal hypertrophy. Vasodilators and catecholamine infusions were discontinued, and CPB was reinstated at a flow of 2.2 l · min<sup>-1</sup> · m<sup>2</sup><sup>-1</sup>.

After aortotomy, inspection revealed that the aortic valve was normal; however, the septum was markedly hypertrophied and exhibited a small area of fibrosis. A part of the septum (1 × 1 × 5 cm) was excised to relieve the left ventricular outflow obstruction.<sup>2</sup> Without

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