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Anesthesia for a Patient with Kufs' Disease

RAY J. DEFALQUE, M.D.*

Kufs', or Kufs-Boehme's, disease was described by Kufs in 1935.[1] It is the adult form of neuronal ceroid lipofuscinosis, an autosomal, usually recessive, inherited disease first described by Batten in children in 1903.[1] Because of its clinical similarity with Tay-Sachs disease, Batten called it "amaurotic familial idiocy," but blindness does not occur in Kufs' form. The disease is due probably to a defect or deficiency in a still unknown enzyme, leading to accumulation of fluorescent lipopigments (ceroid and lipofuscin) in the cytoplasm of neurons and astrocytes of the cerebral cortex, the basal ganglia, and the substantia nigra, and eventually causing cellular degeneration.

Various classifications have been proposed for the several clinical subgroups of the disease. The most recent[1] describes four forms: infantile, late infantile, juvenile, and adult (Kufs'). Kufs' disease appears in early adulthood and usually is progressively fatal over 7–10 yr. It is characterized by variable dementia, severe and drug-resistant grand mal and myoclonic seizures, marked cerebellar ataxia, extrapyramidal spasticity and choreoathetosis, and mild systolic and diastolic hypertension.

The lipopigments also appear in the cytoplasm of the lymphocytes and the cells of the skin and the digestive tract. The diagnosis is confirmed by electron microscopic findings of these pigments in the lymphocytes and cutaneous and mucosal cells biopsies. The treatment is symptomatic and supportive.

CASE REPORT

A 38-yr-old man with Kufs' disease since 1983 was scheduled for exploratory celiotomy after a few days of nausea, constipation, and right upper quadrant rigidity and tenderness. His past history described slowly progressive dementia, severe grand mal and myoclonic seizures, cerebellar ataxia, hypertension, and allergy to phenobarbital and penicillin. He was taking clonazepam 0.5 mg bid, valproic acid 1 g tid, phenytoin 300 mg hs, thioridazine 50 mg bid, and enalapril 5 mg qAM.

He appeared ill-nourished (weight 64.5 kg, height 175 cm) and was uncommunicative and quiet. His ataxia confined him to a wheel chair. The small muscles of his face and hands showed involuntary movements; it was unclear whether these were extrapyramidal manifestations of his disease or were thioridazine-induced dyskinesia.[2] He was edentulous and slightly hypotensive (90/60 mmHg) and had abdominal rigidity and tenderness. The rest of his physical examination and his vital signs, x-rays, ECG, and laboratory tests were normal.

The patient received 300 mg phenytoin the evening before surgery and 0.5 mg clonazepam, 1 g valproic acid, 50 mg thioridazine, and 150 mg ranitidine HCl 2 h before induction. He became agitated during the insertion of an intravenous catheter and was calmed with 2 mg iv midazolam given over 5 min. Three minutes after the patient received 3 mg *d*-tubocurarine iv and while he was breathing oxygen, a rapid-sequence induction was performed with the use of cricoid pressure, 200 mg etomidate, and 100 mg succinylcholine iv. After tracheal intubation, anesthesia was maintained with 1.5 l N_2O–1.5 l 0_2 per min and 0.5–1% inspired isoflurane (mass spectrometer). Four milligrams vecuronium iv was given at the return of the second twitch of the train-of-four. Blood pressure remained at 100/60 mmHg and pulse at 70–80 beats per min throughout a 2.5-h exploratory laparotomy and appendectomy. Administration of the anesthetics was discontinued and 0.6 mg glycopyrrolate iv and 50 mg edrophonium Cl iv were given at the closure of the anterior rectus fascia.

At the completion of surgery the patient was breathing spontaneously with a 350–400-ml tidal volume, 100% 0_2 saturation on room air, with no spectrometer-detectable isoflurane in the expired breaths, and with sustained tetanus (20 Hz, 5 s) All stimuli, however, failed to awaken him or to cause a response to pain. He was taken to the recovery room breathing 60% 0_2 *via* a T-tube attached to the endotracheal tube. Ninety minutes later he abruptly awakened and his trachea was extubated. He recovered uneventfully and was discharged 5 days after his surgery.

DISCUSSION

It was decided to treat this patient as an uncooperative, cachectic subject at risk for perioperative aspiration and to avoid drugs potentially dangerous in epilepsy, dementia, and parkinsonism. Although phenothiazines are contraindicated in Parkinson's disease,[3] this patient received thioridazine preoperatively to keep him calm and cooperative. Preoperative metoclopramide HCl was omitted since it antagonizes dopamine and may exaggerate the parkinsonian symptoms.[4] Droperidol was not considered, for the same reasons.[5] Fentanyl[6] and sufentanil[7] seemed unwise choices in this severe epileptic because of their epileptogenic potential. The selection of a rapid-induction agent caused a dilemma. Ultrashort barbiturates were avoided because of the patient's cachexia, preoperative hypotension, and allergy to phenobarbital. Ketamine seemed contraindicated because of reports of its complications in epilepsy,[8] parkinsonism,[9] and dementia.[10,11] Etomidate was chosen although its use in epileptics is controversial.† Because of its potential for hyperkalemia in Parkinson's disease[12] and ataxia,[13] succinylcholine was a debatable choice but seemed preferable to a nondepolar-

* Professor, Dept of Anesthesiology.

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Address reprint requests to Dr. Defalque: Department of Anesthesia, University of Alabama at Birmingham, Birmingham Veterans Administration Medical Center, Birmingham, Alabama 35233.

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† Glass P, Leiman BC, Reeves JG: Etomidate: What is its present role in anesthesia? Seminars in Anesthesia 7:143–151, 1988

izing drug in this edentulous patient at risk for aspiration and with possible phenytoin-induced resistance to a nondepolarizing muscle relaxant.[13] Isoflurane seemed a good choice for an ill-nourished epileptic.[14]

It is unclear why this patient was slow to awaken despite mass spectrometer indication of little or no residual anesthetic. Temperature, urinary output, electrolytes, and arterial blood gases were normal in the immediate postoperative period, and there was no sign of central nervous system changes once the patient awakened. Prolonged sedation from the preoperative medications is a possibility, although it seems doubtful in consideration of phenytoin-induced liver enzyme stimulation.[15] However, valproic acid has the opposite effect.[15] The patient's preoperative liver function tests were normal.

Although one cannot generalize from a single case, this patient's anesthesia, except for a slow emergence, was uneventful and may provide useful guidelines for those responsible for providing anesthesia to a patient with Kufs' disease.

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Intramyocardial Air Causes Right Ventricular Dysfunction after Repair of a Congenital Heart Defect

WILLIAM J. GREELEY, M.D.,* FRANK H. KERN, M.D.,† ROSS M. UNGERLEIDER, M.D.,‡ JOSEPH A. KISSLO, M.D.§

The intraoperative occurrence of right ventricular dysfunction can complicate the repair of congenital heart defects in children and thereby cause low-output syndrome and difficulty separating from cardiopulmonary bypass. Suggested etiologies for right ventricular dysfunction after surgical repair include: myocardial ischemia due to inadequate protection during bypass; exacerbation of preexisting right ventricular hypertrophy and dilation due to chronic congenital heart defects; right ventriculotomy; pulmonary artery hypertension; and direct damage during surgical repair to the right ventricle or tricuspid valve apparatus.[1] We report a case of intramyocardial air identified intraoperatively by epicardial Doppler echocardiography (echo-Doppler) contributing to right ventricular dysfunction during congenital heart surgery in a child and suggest a management plan.

* Associate Professor of Anesthesiology and Pediatrics; Associate, Pediatric Cardiology.
† Assistant Professor of Anesthesiology; Associate in Pediatrics.
‡ Assistant Professor of Surgery.
§ Professor of Medicine.
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Address reprint requests to Dr. Greeley: Box 3046, Duke University Medical Center, Durham, North Carolina 27710.
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