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LETTER TO THE EDITOR

Anesthetic management for a patient with very-long-chain acyl-coenzyme A dehydrogenase deficiency

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To the Editor:

A 28-year-old woman with the adult form of very-long-chain acyl-coenzyme A dehydrogenase (VLCAD) deficiency was scheduled for hysteroscopic myomectomy. She complained of muscle pain on fasting and hard exercise, which was alleviated by having food or resting. She was taking carnitine and could usually tolerate overnight fasting. So she fasted for 8 h before the operation, and fluid infusion was begun in the operation room with maintenance fluid containing 10 % glucose at 4.5 mg/kg/min. We used diazepam for premedication. General anesthesia was induced with thiopental, remifentanil, and rocuronium and maintained with sevoflurane and remifentanil. A diclofenac suppository was administered for postoperative analgesia. During the surgery, hyperglycemia (254 mg/dl) was treated with insulin (2 units). Lactate was slightly elevated. The operation was completed uneventfully. Neuromuscular blockade was antagonized with sugammadex; she awoke completely and made no complaint of pain, nausea, vomitting or shivering. An infusion containing 5 % glucose was continued until the next morning, with oral feeding being started the afternoon of the same day.

VLCAD is one of the enzymes involved in mitochondrial long-chain fatty acid β-oxidation. A lack of VLCAD activity leads to a shortage of the energy normally derived from long-chain fatty acids [1, 2]. This means that energy is supplied only by glucose, amino acids, or lipids other than very-long-chain and long-chain fatty acids. Catabolic fasting, certain stresses, and an elevated energy demand can induce severe metabolic decompensation [2]. Additionally, the increases in acyl-coenzyme A and acylcarnitine are harmful. The symptoms of VLCAD deficiency are derived from organs/tissues such as the heart, liver, and skeletal muscle, where mitochondrial fatty acid β-oxidation is most marked. The severe form of VLCAD deficiency manifests with cardiomyopathy and has a high mortality rate [1]. Treatment of VLCAD deficiency is directed at preventing catabolic situations by the avoidance of prolonged fasting, by avoiding emotional or physical stress, and by dietary restriction of long-chain fatty acids. In the present patient, our aims were: (1) not to elevate plasma free fatty acids, (2) not to increase energy demand, and (3) to give sufficient glucose. We chose general anesthesia to avoid the physical and/or psychological stresses that could be induced by a possibly inadequate level of spinal anesthesia. Some cases have been reported in which propofol or sevoflurane was used safely in patients with VLCAD deficiency [3]. However, propofol contains long-chain fatty acids, and we therefore avoided using this agent. Some stresses (anxiety, shivering, nausea, and vomiting) can increase energy demand, and a previous report has recommended glucose, at 6 mg/kg/min, for perioperative use in infants with VLCAD deficiency; as well, it was suggested that the fasting period should not exceed 4 h in children with this condition [4]. However, it is not clear how much glucose is necessary during an operation in adult patients with VLCAD deficiency. Because glucose demand may decrease with age and because myopathic symptoms did not occur and the creatine kinase level was not elevated just after the operation in our patient, we think that the intraoperative glucose infusion she received was adequate. However, creatine kinase was elevated on postoperative day 2, and we speculate that the postoperative glucose infusion was not

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sufficient or that she had needed an earlier resumption of oral food intake. Because rhabdomyolysis may occur under conditions of normoglycemia in patients with VLCAD deficiency [5], careful postoperative monitoring of glucose, creatine kinase, and the clinical condition is desirable in these patients.

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