Can Inhaled Induction Be Considered As Safe In Patients With Mc Ardles Disease?

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My opinion

McArdle’s disease (myophosphorylase deficiency) is a glycogen storage disease, which can present with symptoms as exercise intolerance, muscle cramps, fatigue, rhabdomyolysis, myoglobinuria and has been associated with the possible risks of acute renal failure and malignant hyperthermia [1,2]. Isaacs et al. suggested that all patients with McArdle’s disease should be tested for malignant hyperthermia (MH) prior to surgery [3]. Benca and Hogan [4] suggested that inhaled induction of anaesthesia with “the newer potent inhaled anaesthetics sevoflurane or desflurane” in order to place an IV catheter may be an option in patients with “rare enzyme defects” (like McArdle’s disease). They stated that the evidence for an association between McArdle’s disease and malignant hyperthermia (MH) is weak although some patients have a positive caffeine halothane contracture test (CHCT) [4]. No cases of malignant hyperthermia in patients with McArdle’s disease have been described so far, but one patient had rhabdomyolysis and hyperthermia intraoperatively [5]. Although the authors of this case report concluded that the rhabdomyolysis and hyperthermia were linked to a reaction to protamine they stated:

“The presence of McArdle’s disease may have rendered this patient more susceptible to skeletal muscle injury when increased energy demand ensued, thus resulting in massive rhabdomyolysis and renal failure.” [5]

Patients with McArdle’s disease might react with muscle cramps or rhabdomyolysis, myoglobinuria to different causes and have a risk for developing renal failure due to rhabdomyolysis. Therefore one should avoid muscle ischemia as e.g. caused by tourniquets and try to prevent shivering [2]. An own study included a patient with McArdle’s disease and a positive CHCT. This patient had had an episode of tachycardia and hypotonia under general anesthesia with the use of both succinyl, halothane and isoflurane and did not develop MH [2].

At present it is unclear if a positive CHCT test result in McArdle’s disease is associated with the risk for MH or if it is just a reaction of a muscle running out of fuel.

Until this question is answered one should treat patients with McArdle’s disease as patients with the risk of developing MH. In general whenever giving anaesthesia to patients with McArdle’s disease the anaesthetist should avoid MH-trigger substances and consider the recommendations given by me, Mohr and Ræder if not otherwise contraindicated [2]. The evidence for a connection between McArdle’s disease and MH is weak and the risk of developing MH is low for patients with McArdle’s disease. Nevertheless we do not know yet how to interpret a positive test result in the CHCT in McArdle patients. Taking into account the present state of scientific knowledge inhaled induction cannot be recommended as a safe option for patients with McArdle’s disease.

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