Liver transplantation in a child with osteogenesis imperfecta

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We read with great interest the case report by Lee et al. [1], of the anesthetic management of an infant with osteogenesis imperfecta (OI) and progressive familial intrahepatic cholestasis, who was undergoing orthotopic liver transplantation (OLT). This case was indeed challenging, but we would like to add the following comments.

First, OI is only weakly associated with malignant hyperthermia, if at all. [2] Cases of intra- or post-operative hyperthermia have indeed been reported, and are thought to be caused by the chronic basal hypermetabolism associated with abnormalities of the collagen structure, thereby resulting in OI. However, a large, recently published case series has demonstrated that the risk of hyperthermia is much lower than was initially assumed. [3] Moreover, over a number of years, thousands of children with OI have received halogenated-based anesthesia with no complications: no case of anesthesia-induced rhabdomyolysis has been observed.

Second, the use of propofol-based total intravenous anesthesia, during OLT, is a pharmacologic challenge: initial hepatic function, major fluid shifts and the presence of an anhepatic phase render adaptation of the dosage of propofol to the patient's needs, without measuring its effect on the brain, almost impossible. Only one previous case, of an adult who underwent two OLTs, has been published. [4] In the pediatric case described, liver function was probably initially normal but, as the authors discuss, BIS is unreliable in children of less than 2 years of age: it would therefore be instructive to demonstrate how the infusion rates of propofol and remifentanyl were adapted to during

surgery, particularly during the anhepatic phase and the initial phase of hepatic reperfusion. The use of a 2% propofol infusion represented a prudent decision to avoid overloading the newly transplanted liver with triglycerides.

Third, regarding the prevention of propofol-infusion-syndrome (PRIS), it is recommended that providing > 4 mg/kg/h propofol be avoided, and further that co-administration of sufficient glucose (6 mg/kg/min) might protect against mitochondrial dysfunction [5]: higher doses of propofol (6 mg/kg/h) were apparently used in this case and no data regarding glucose are provided. Using the model of Short, and taking into account both the anhepatic phase and the presence of extrahepatic metabolism, the addition of ketamine or midazolam could have been considered. The early diagnosis of PRIS is difficult during OLT, because the initial biological sign in healthy patients is unexplained lactic acidosis. Fortunately, lactate levels remained low in the presently discussed case. It would be instructive to know the blood lactate threshold used to diagnose PRIS, and the alternative strategy in place if PRIS had occurred.

In closing, there are many possible causes of hyperthermia following OLT, including infection, transfusion, rebound hyperthermia following intraoperative hypothermia, iatrogenesis, and malignant hyperthermia (MH). Fever represents a late sign of MH: other indicators are generally present at the time of initial occurrence, such as hypercarbia, hyperkalemia, dysrhythmias, and dark urine (myoglobinuria). In this context, the blood gases observed upon arrival at the PICU were more concerning, in terms of indicating a possible MH crisis, compared with those measured at the time of fever.

The authors should be congratulated for drawing attention to the possible pharmacologic effects of non-anesthetic drugs (namely to those contained in the liver conservation solution) administered during an OLT.

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References

1. Lee J, Ryu HG, Kim A, Yoo S, Shin SY, Kang SH, et al. Anesthetic management of an 8-month-old infant with osteogenesis imperfecta undergoing liver transplantation: a case report. *Korean J Anesthesiol* 2014; 66: 472-5.
2. Benca J, Hogan K. Malignant hyperthermia, coexisting disorders, and enzymopathies: risks and management options. *Anesth Analg* 2009; 109: 1049-53.
3. Bojanic K, Kivela JE, Gurrieri C, Deutsch E, Flick R, Sprung J, et al. Perioperative course and intraoperative temperatures in patients with osteogenesis imperfecta. *Eur J Anaesthesiol* 2011; 28: 370-5.
4. Schmidt A, Tzanova I, Köbler AM, Wissner GJ, Kentner R. Orthotopic liver transplantation in a malignant hyperthermia susceptible patient. *J Clin Anesth* 2005; 17: 558-61.
5. Kam PC, Cardone D. Propofol infusion syndrome. *Anaesthesia* 2007; 62: 690-701.