Glutaric aciduria type 1: anaesthetic management of an adult patient for elective laparoscopic cholecystectomy

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Background

Glutaric aciduria type 1 (GA1) is a rare hereditary metabolic disorder (prevalence of 1 in 100 000 newborns).

Untreated, approximately 90% of patients will develop neurological disease following an acute encephalopathic crisis often precipitated by gastroenteritis, intercurrent febrile illness, immunization or **surgical intervention**.

Case Report

- ♂, 20-year-old (weight 45 Kg, height 165 cm) submitted to elective laparoscopic cholecystectomy
- Personal history: GA1 diagnosed at 1-year-old treated with low protein diet, riboflavin and carnitine. No acute metabolic disturbances in the past year
- Clinical examination: psychomotor delay, dystonia and dysarthria. Normal routine laboratory tests

Discussion

Anticipate anaesthetic risks in GA1 patients:

- Dehydration
- Hyperthermia and catabolic state
- Possibility of pulmonary aspiration
- Prolonged responses to nondepolarizing muscle relaxants
- Hyperkalemic responses to succinylcholine

Main question: volatile anaesthetic agents - yes or not?

Against the use of volatile anesthetics → possible association between mitochondrial dysfunction and malignant hyperthermia? More recent literature rules out episodes of malignant hyperthermia and other intraoperative events attributable to the general anesthesia with sevoflurane in paediatric patients with mitochondrial disease.

Against propofol → lipid overload and inhibition oxidative phosphorylation, carnitine palmitoyltransferase transport of long-chain fatty acids, and β -oxidation of fatty acid in mitochondria.

Although the described case was successful, the anaesthetic management of patients with metabolic disorders like GA1 remains challenging and more clinical data are necessary to improve these patients outcomes.

Anaesthetic Management

- Fluid therapy: 5% dextrose in saline
- Midazolam 1mg IV before entering the OR
- Standard monitoring; active warming
- Induction: fentanyl 250 mcg + **thiopentone 125 mg** + rocuronium 20 mg (IV)
- Mixture of air-oxygen (FiO_2 0,5) + **sevoflurane**
- PONV: ondansetron 4 mg (IV)
- Analgesia: paracetamol 500 mg (IV)
- To reverse NMB: sugammadex 200mg (IV)
- Surgery successfully completed in 40 min
- Oral intake resume after 4 h
- The post operative period was uneventful

References:

K. Stefan et al; Diagnosis and management of glutaric aciduria type I – revised recommendations; J Inherit Metab Dis (2011) 34:677–694; H.P. Joaquin et al; Anesthetic management in two siblings with glutaric aciduria type 1; Pediatric Anesthesia (2006) 16: 188–191; W.N. Teng et al; Anesthetic management of comprehensive dental restoration in a child with glutaric aciduria type 1 using volatile sevoflurane; Journal of the Chinese Medical Association 77 (2014) 548-551.