When is the benefit greater than the risk?

Authors: Adriana Rodrigues; Carla Hipólito; Cristina Gomes.

Institution: Hospital de Braga, Braga, Portugal

Background

Pseudohypoaldosteronism type I (PHA I) is characterized by a mineralocorticoid resistance caused by an inactivating mutation of the gene encoding ENaC. It can lead to salt wasting crises and severe hyperkalemia causing life-threatening cardiac arrhythmias. The systemic form of the disease requires a life-long salt supplementation¹.

PHA type I

Preoperative assessment

- ♂, 6y, 16,5 Kg
- Systemic PHA 1 treated with 240mL NaCl 20%
- Tonsillectomy, adenoidectomy and myringotomy

Intraoperative management

- Na⁺ at admission: 129mmol/L
- Administered 50mL NaCl 20% after induction
- Metoclopramide, dexamethasone, ondansetron
- Postoperative Na⁺: 145 mmol/L

Postoperative care

- Surgery lasted 90 minutes
- Extubated uneventfully
- Recovery in a pediatric intermediate care unit
- Discharged home two days after, asymptomatic

Discussion

PHA I is a rare disease that places patient at risk of severe dehydration and ionic imbalance². The ionic variations in this child are difficult to predict, so we decided to prompt correct the imbalances administering NaCl through a gastric tube in order to prevent severe hyponatremia and hyperkalemia. It would be impossible to perform this amount of sodium supplementation IV due to the large volume of fluid needed in short time. Although there was the possibility of supplementation at the end of surgery it would still exist possibility of regurgitation and aspiration. We are aware of some conditions that we could optimize before surgery, namely oral ingestion of liquids until 2 hours before surgery and early onset of fluid therapy. We inadvertently overcorrect the sodium level to which, may contribute intraoperative use of dexamethasone and hydrocortisone.

Learning points

Anaesthesiologist always must balance the risks and the benefits of his attitudes. Maybe sometimes it worth it to take the risks.

References:

- ¹) Tajima T. et al. Clinical features and molecular basis of pseudohypoaldosteronism type 1. Clin Pediatr Endocrinol 2017; 26(3), 109–117.
- ²) Nur N. et al. Systemic Pseudohypoaldosteronism Type I: A Case Report and Review of the Literature. Case Reports in Pediatrics (2017); 1-6.