Anesthesia for the *Pediatric* Patient with Hypokalemic Periodic Paralysis

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Quick Reference Guide

Hypokalemic Periodic Paralysis (HypoKPP) is an autosomal dominant disease with possible mutations or variants at CACNA1S, SCN4A, or RyR1.[1,2,3] These patients experience periods of flaccid muscle paralysis based on decreased serum potassium (K^+) levels.[2,4,5] The Malignant Hyperthermia Association website lists HypoKPP as a condition associated with malignant hyperthermia (MH).[1,4,6] With low prevalence in the general population, most treatment comes from anecdotal reports and not all patients respond alike.[4,6,7]

Daily Prevention: HypoKPP patients need a low sodium and carbohydrate diet, avoidance of attack triggers noted below, and no alcohol.[4,8,9] Pharmacological interventions may include a K^+ salt, carbonic anhydrase inhibitor, and a K^+ sparing diuretic.[4,8]

Emergency Treatment: Oral K^+ is preferred if the patient can swallow.[8,10] K^+ may also be given via an oral/nasogastric tube or intravenously. The intravenous (IV) potassium chloride (KCl) must not be mixed in a dextrose solution.[10] The usual pediatric rate of KCl infusion should not exceed 10 mEq per hour, but heart dysrhythmias and respiratory compromise may require temporary higher infusion rates.[10] **Avoid aggressive K^+ replacement as HypoKPP is a shift in K^+, not a loss of K^+.**[4,8,10] Safe practice includes EKG monitoring with frequent serum K^+ level checks.[4] Patients with ongoing paralysis episodes have been shown to benefit from an immediate slow infusion of KCl.[11]

Which HypoKPP patient may be susceptible to a MH crisis? Any patient with a personal or close family history of a MH-like episode**, *OR*** any patient with mutant variants of RyR1, CACNA1S, ***OR*** if unknown genotype, even if no past anesthesia problems.[2,9,12,13,14,15] In the general population, children < 15 years old comprise 52.1% of all MH reactions.[15] Most patients/caretakers know their genetic mutation.

Pediatric Anesthetic Plan

Standard of Care:	Core temperature > 36°C, avoid dehydration, keep serum K^+ at or above patient's target level if known, otherwise use upper normal level (i.e., 5 mEq/L)[16] and avoid triggers.[5,8,9,14,16,17,18,19]
Avoid Attack Triggers:	Serum K^+ level below patient's target level, stress, cold, excitement, fear, pain, fasting, Na^+ and glucose loads via IV or oral intake, **steroids, epinephrine**, **succinylcholine**, insulin, hyperglycemia, metabolic or respiratory alkalosis, certain antibiotics, and anesthesia. [4,5,8,9,12,14,16,18,19]
Regional Anesthesia:	Preferred: Spinal, epidural, regional nerve block, and local infiltration with appropriate sedation and monitoring. Case reports use normal dosing ranges.[16,18,19,20,21]
Local Anesthetics:	May use **plain** lidocaine, bupivacaine, or ropivacaine.[5] Lidocaine is not always effective.[5] **No epinephrine.**[5,20]
Pre-op Testing / Consultations:	Serum Na^+, K^+, Ca^{++}, Mg^{++}, EKG, and possibly PFT. Consult patient primary care physician and specialists.[5,14,18]
Preparations:	Point of Care K^+ monitor, warm the OR, patient warming device, fluid warmer, and pump for KCl drip.[5,14,16]
Intra-op Monitoring:	Core temperature, point of care K^+ levels, nerve stimulation, and end-tidal CO_2 ($ETCO_2$).[5,16,18,19]

Anesthesia Process for HypoKPP Patient

Preoperative	EMLA cream may help IV start & electrolyte monitoring access.[16,17,22] Sedate using benzodiazepine like midazolam or diazepam.[5,16,17,18,19] Watch for respiratory depression.[5,6,18] Preferred fluid is lactated ringers (No dextrose & limit Na^+ load).[5,9,12,14,17,18] Correct electrolyte levels and verify that ***serum K+ is at patient's target level***.[5,14,16,17]
MH Susceptible Anesthesia Plan	Start IV and use propofol for induction. Maintain with a TIVA using a propofol drip.[16,17,23] ***No succinylcholine*** or anesthesia gases except nitrous oxide.[5,13,24] Avoid etomidate→ decreases K^+.[22] Can use remifentanil.[16,17]
NOT **MH Susceptible Anesthesia Plan**	IV induction with propofol.[16,17] (Avoid etomidate→ decreases K^+)[22] ***OR*** use inhalation induction with sevoflurane and/or nitrous oxide.[18] Don't use halothane[15] or desflurane→ upper airway events.[22] ***No succinylcholine***.[4,5,12,18] Maintain with TIVA (propofol)[16,17,23] ***OR*** inhalation gases→ sevoflurane or isoflurane.[18,19] Can use remifentanil.[16,17]
Maintenance for both plans	Keep $ETCO_2$ around 40 mmHg for hypercarbia to maintain acidosis.[9,12,16,17,18,19] Check serum K^+ with any change in patient condition, vital signs, or EKG and titrate K^+ to patient target level[5,16,18,19,21]. Keep core temperature >36°C, good pain control, minimal muscle relaxation, and avoid dehydration.[5,14,19,21] A slow KCl IV infusion may help.[11,19]
Paralysis	***No succinylcholine***.[4,5,12,18] Short-acting non-depolarizing muscle relaxant-start at 10-20% normal dose.[4,6,9,12,14,17,18,19]
Pain Management	**Regional Anesthesia works best!** [16,18,19,20,21] Weight-based dosing of acetaminophen & ketorolac.[17,22] Opioids given by incremental low doses to the desired effect.[5,16,17] Monitor for chest wall rigidity and respiratory drive.[5,22]
Antiemetics	No steroids→ attack trigger.[5,8,9] No literature was found on antiemetic use with HypoKPP patients.
Emergence	Use short acting nondepolarizing muscle relaxant - Anticholinesterases are not recommended.[18]
Recovery	Consider ICU. Close K^+ control, trigger avoidance, electrolyte management (i.e., post-op vomiting→ may decrease Mg^{++}→ makes K^+ replacement ineffective) to prevent a paralytic attack.[5,6,9,16,17] Carefully manage pain (a paralytic episode does not decrease pain level)[16,18,19,21]. Post-op diet must be low Na+ and low carbohydrate.[8,9] Early family (caretaker) involvement may help identify paralytic attacks.[14] Resume patient's daily medications ASAP.[18]

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This document gives anesthesia providers basic guidelines designed to increase the safe anesthesia care for this patient population. Being a rare disease, most of the information is based on the lower level of the evidence hierarchy and does not consider comorbid conditions. Feedback and comments may be forwarded to PeriodicParalysisResearch@gmail.com

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