

Anesthesia for the Adult 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 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 infusion rate of KCl is 10 mEq/hour, but not to exceed 20-25 mEq/hour as 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 continuous EKG monitoring with frequent serum **K⁺** level checks.⁴ Patients with ongoing paralysis episodes benefit from 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}

Pregnancy: No contraindications for normal childbirth.⁶ Labor epidurals work well for pain and anxiety.^{16,17} Spinals are beneficial for a Cesarean Section.^{5,18} No epinephrine.^{5,17} Control glucose, **K⁺** and patient temperature.^{4,17} Use guidelines below for general anesthesia.

Adult 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) ¹⁸ and avoid triggers. ^{5,8,9,14,16,18,19,20}
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. ^{4,5,8,9,12,14,16,18,20}
Regional Anesthesia:	Preferred Anesthesia Choice: Spinal, epidural, regional nerve block, and local infiltration with appropriate sedation and monitoring. Case reports use normal dosing ranges. ^{16,17,18,20,21}
Local Anesthetics:	May use plain lidocaine, bupivacaine, or ropivacaine. ⁵ Lidocaine is not always effective. ⁵ No epinephrine. ^{5,17}
Pre-op Testing / Consultations:	Serum Na⁺ , K⁺ , Ca⁺⁺ , and Mg⁺⁺ , EKG, and possibly PFT. Consult patient primary care physician and specialists. ^{5,14,16}
Preparations:	Point of Care K⁺ monitor, warm the OR, patient warming device, fluid warmer, and pump for KCl drip. ^{5,14,18}
Intra-op Monitoring:	Core temperature, point of care K⁺ levels, nerve stimulation, and end tidal CO₂ (ETCO ₂). ^{5,16,18,20}

Anesthesia Process for HypoKPP Patient

Preoperative	IV start and obtain access for electrolyte monitoring. ^{18,19,22} Sedate using benzodiazepine (i.e., midazolam or diazepam). ^{5,16,18,19,20} Watch for respiratory depression. ^{5,6,16} Preferred fluid is lactated ringers (No dextrose & limit Na⁺ load). ^{5,9,12,14,16,19} Correct electrolyte levels and verify that serum K⁺ is at patient's target level. ^{5,14,18,19}
MH Susceptible Anesthesia Plan	IV induction with propofol. Maintain with a TIVA using a propofol drip. ^{18,19} No succinylcholine or anesthesia gases except nitrous oxide. ^{5,13,23} Avoid etomidate → decreases K⁺ . ²² Can use remifentanyl. ^{18,19}
NOT MH Susceptible Anesthesia Plan	IV induction with propofol. ^{18,19} (Avoid etomidate → decreases K⁺). ²² No succinylcholine. ^{4,5,12,16} Maintain with TIVA (propofol) ^{18,19} OR inhalation gases → sevoflurane or isoflurane preferred. ^{16,20} Avoid desflurane to minimize upper airway events. ²² Can use remifentanyl. ^{18,19}
Maintenance for both plans	Keep ETCO ₂ around 40 mmHg for hypercarbia to maintain acidosis. ^{9,12,16,18,19,20} 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, & avoid dehydration. ^{5,14,20,21} A slow KCl IV infusion may help. ^{11,20}
Paralysis	No succinylcholine. ^{4,5,12,16} Short-acting non-depolarizing muscle relaxant-start at 10-20% normal dose. ^{4,6,9,12,14,16,19,20}
Pain Management	Regional Anesthesia works best! ^{16,17,18,20,21} Weight based dosing of acetaminophen & ketorolac. ^{19,22} Opioids given by incremental low doses to the desired effect. ^{5,18,19} Monitor for chest wall rigidity and respiratory drive. ^{5,22}
Antiemetics	No steroids → attack trigger. ^{5,8,9} No literature found on antiemetic use with HypoKPP patients.
Emergence	Use short acting nondepolarizing muscle relaxant - Anticholinesterases are not recommended. ¹⁶
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. ^{5,6,9,18,19} Carefully manage pain (a paralytic episode does not decrease pain level) ^{16,18,20,21} . 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. ¹⁶

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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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