Anaesthesia
pearls and pitfalls in periodic paralysis

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General remarks

• Periodic Paralyses are rare diseases
• Physicians are often not familiar with PP
• Patients should be better informed than their doctors
• Anesthesiologists are specialists for anesthesia, not for rare diseases
ABSTRACT: Patients with neuromuscular disease pose many anesthetic challenges and are at greater risk for perioperative complications, including respiratory or cardiovascular dysfunction and pulmonary aspiration. Therefore, these patients require special precautions, including interdisciplinary collaboration among primary care physicians, neurologists, surgeons, and anesthesiologists. Preoperative optimization of comorbid conditions is crucial, as patients may have adverse response to neuromuscular blocking drugs and the reversal drugs (e.g., neostigmine and edrophonium) should be used with caution and titrated based on effective neuromuscular monitoring. Drugs that potentiate neuromuscular blocking drugs should also be avoided or their doses limited if possible. The risk of malignant hyperthermia in certain neuromuscular diseases mandates avoidance of triggering agents such as succinylcholine and inhaled anesthetics. Patients with neuromuscular disease may also be sensitive to sedative-hypnotics and opioids, which should be used judiciously. Finally, the postoperative period requires close monitoring due to increased risk of postoperative cardiorespiratory dysfunction.


There is no role for routine preoperative testing. Patients’ functional status and the type of surgery should dictate preoperative testing. Certain drugs such as corticosteroids, aminoglycosides, vancomycin, quinidines, ester-type local anesthetics, furosemide, calcium channel blockers, and beta-blockers can amplify neuromuscular blockade. Also, preoperative plasmapheresis can prolong the effects of succinylcholine, mivacurium, remifentanil, and esmolol, as they are metabolized by plasma cholinesterase, which may be reduced by plasmapheresis.
PP and general anaesthesia
main problems

• HyperPP and PC
  – Sux and $K^+$ cause severe muscle stiffness
    • Sux depolarises and then inactivates voltage-gated $Na^+$-channels

• HypoPP
  – Stress causes hypokalemia and severe flaccid muscle weakness

• PC and PP
  – Cooling causes flaccid muscle weakness
PP and general anesthesia

Succinylcholine & K+ cause severe muscle stiffness (can impair intubation and ventilation)

On the right:
Succinylcholine-induced contraction of an excised myotonic muscle bundle
Pre-operative considerations

• Is surgery really necessary?
• Pre-Surgery-examination
• Full neurological exam
• ECG
• Chest X-Ray
• Echokardiography
• Pulmonary function tests with body plethysmography
• Arterial blood gas analysis
• Serum: Na^+, K^+, Cl^−, Mg^{2+}, Ca^{2+}, CK, Myoglobin
Pre-Medication

• Benzodiazepines possible
• No NaCl 0.9%
• No pure Glucose infusions
• Stop Statins (lower Cl⁻-conductance (similar extent as in Cl⁻-channel myotonia))
• Consider Beta-Blockers to reduce perioperative stress
During surgery

• Invasive monitoring enables repetitive blood gas analysis
• Temperature monitoring – avoid hypothermia
• Use short acting intravenous drug (propofol + opioids)
• No suxamethonium, no depolarising muscle relaxants, especially in myotonia fluctuans
• Use clonidine and nefopam to reduce shivering
• Consider postoperative ICU
Complications

• Rhabdomyolysis (after suxamethonium): not a special problem in PP, because the muscle fibers are not denervated

• No special cardiac complications in PP (except ATS, except severe hypokalemia)

• Respiratory distress: not a special problem in PP, because involvement of the respiratory muscles is rare (however respiratory depressive drugs may lead to decompensation)
Complications

- Myotonic reactions (hyperPP) can be induced by depolarising agents, $K^+$, anticholinesterases, and opioids
- Suxamethonium: myotonia of the jaw (masseter)
- Myotonic reactions aggravated by alteration of serum osmolarity, pH, ambient temperature, by hypothermia-induced shivering
- TX: lidocain, mexiletin
Complications

• Masseter spasms, muscle spasms
• DD Myotonia, increased sarcoplasmatic Ca$^{2+}$ release (MH, slowed reuptake Brody-disease)
• TX: Dantrolene (Ca$^{2+}$)
• Hyperthermia: not a problem in PP (myotonic hyperthermia in hyperPP)
• **Hypothermia**
• Hypothermia increases the sensitivity of muscle to depolarising and non-depolarising muscle relaxants, potentially aggravating rhabdomyolysis or myotonic reactions. Hypothermia potentiates dysrhythmias in the predisposed patient, promotes bleeding, alters the haemoglobin dissociation curve
General anesthesia and HypoPP

Use bear hugger

Warmed operating room

Area of operation to be kept wet with warmed solutions

K kept in the high normal range

No pure NaCl or glucose infusions

Operation-induced stress to be reduced by Beta-Blocker

No Succinylcholine as relaxant (like for MH individuals)
Local anesthesia and HypoPP

No Epinephrine (used for keeping anesthetic localized)

Avoid Lidocaine (can precipitate weakness spell and is not effective in some individuals; in contrast, Ropivacaine is effective in these individuals)
General anesthesia and HyperPP

staff aware of the diagnosis of hyperkalemic PP

Warmed operating room

Area of operation to be kept wet with warmed solutions

After recovering from general anesthesia, patients may be paralyzed for hours

Opioids or depolarizing agents can precipitate a myotonic reaction that may interfere with intubation and ventilation

Modification of the induction sequence
Prevention of carbohydrate depletion

No muscle relaxants
No Succinylcholine as relaxant (like for MH individuals)
Essentials HypoPP

• Operation-induced stress leads to $K^+$ uptake into muscle via release of catecholamines, insulin, and other hormones.
• The resulting hypokalemia, potentially worsened by sodium chloride infusions, as well as mild hypothermia can induce a paralytic attack.
• Keep the patients warm
• Keep serum $K^+$ at high level
• avoid hyperglycaemia
Essentials HyperPP

- inhalational induction
- Respiratory distress: weakness aggravated by drugs that depress respiration and by the hypothermia (Paramyotonia congenita and hyperPP)
- Preventive therapy before surgery
- maintain a normal body temperature
- keep serum $K^+$ at low level
- avoid hypoglycaemia
Summary PP

• No depolarising muscle relaxants (esp. Myotonia fluctuans)
• Induction with $O_2$, Thiopental, 2x ED95 of a nondepolarising relaxant, intubation
• hyperPP: inhalational induction
• Respiratory distress: weakness aggravated by drugs that depress respiration and by the hypothermia (Paramyotonia congenita and hyperPP)
• Preventive therapy before surgery, maintaining a normal body temperature and keeping serum potassium at low level and avoiding hypoglycaemia
• Hypo-PP. Operation-induced stress leads to K+ uptake into muscle via release of catecholamines, insulin, and other hormones. The resulting hypokalaemia, potentially worsened by sodium chloride infusions, as well as mild hypothermia can induce a paralytic attack
  
HypoPP Keeping the patients warm and serum K+ at high level and avoiding hyperglycaemia are essential measures in preventing such attacks
• Careful monitoring of pre-existing QT prolongation during and after anaesthesia is a must
• Regional anaesthesia whenever feasible seems to be preferred despite its well-documented consequence of hypokalaemia
• Generalized muscle spasm: myotonic crisis (not MH)