

Biannual Meeting of the PPA Orlando, FL, 2013

Anaesthesia pearls and pitfalls in periodic paralysis

Department of Neurology
Military Hospital Ulm
Germany



Frank Weber



Frank Lehmann-Horn, Senior
Research Professor

Gemeinnützige Hertie-Stiftung 

General remarks

- Periodic Paralyzes 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

NEUROMUSCULAR DISEASE AND ANESTHESIA

ALAN ROMERO, MD and GIRISH P. JOSHI, MD, FFARCSI

University of Texas Southwestern Medical Center, 5323 Harry Hines Boulevard, Dallas, TX 75390, USA

Accepted 11 February 2013

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 communication between primary care physicians, neurologists, anesthesiologists, and anesthesiologists. Preoperative optimization of comorbid conditions is essential. Patients may have adverse response to neuromuscular blocking drugs and the reversal drugs (e.g., neostigmine) should be used with caution and titrated based on clinical response. Intraoperative 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.

Muscle Nerve 48: 451–460, 2013

PP not mentioned!

(e.g., pulmonary aspiration), and gastroin-
testinal dysfunction. Optimization of these comorbidities is im-
portant. Patients require special precautions to avoid perioperative complications and
improve outcome.

There is no role for routine preoperative test-
ing. Patients' functional status and the type of sur-
gery should dictate preoperative testing.⁵ Certain
drugs such as corticosteroids, aminoglycosides, van-
comycin, 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, remifenta-
nil, and esmolol, as they are metabolized by plasma
cholinesterase, which may be reduced by plasma-
pheresis.² Certain neuromuscular diseases have a

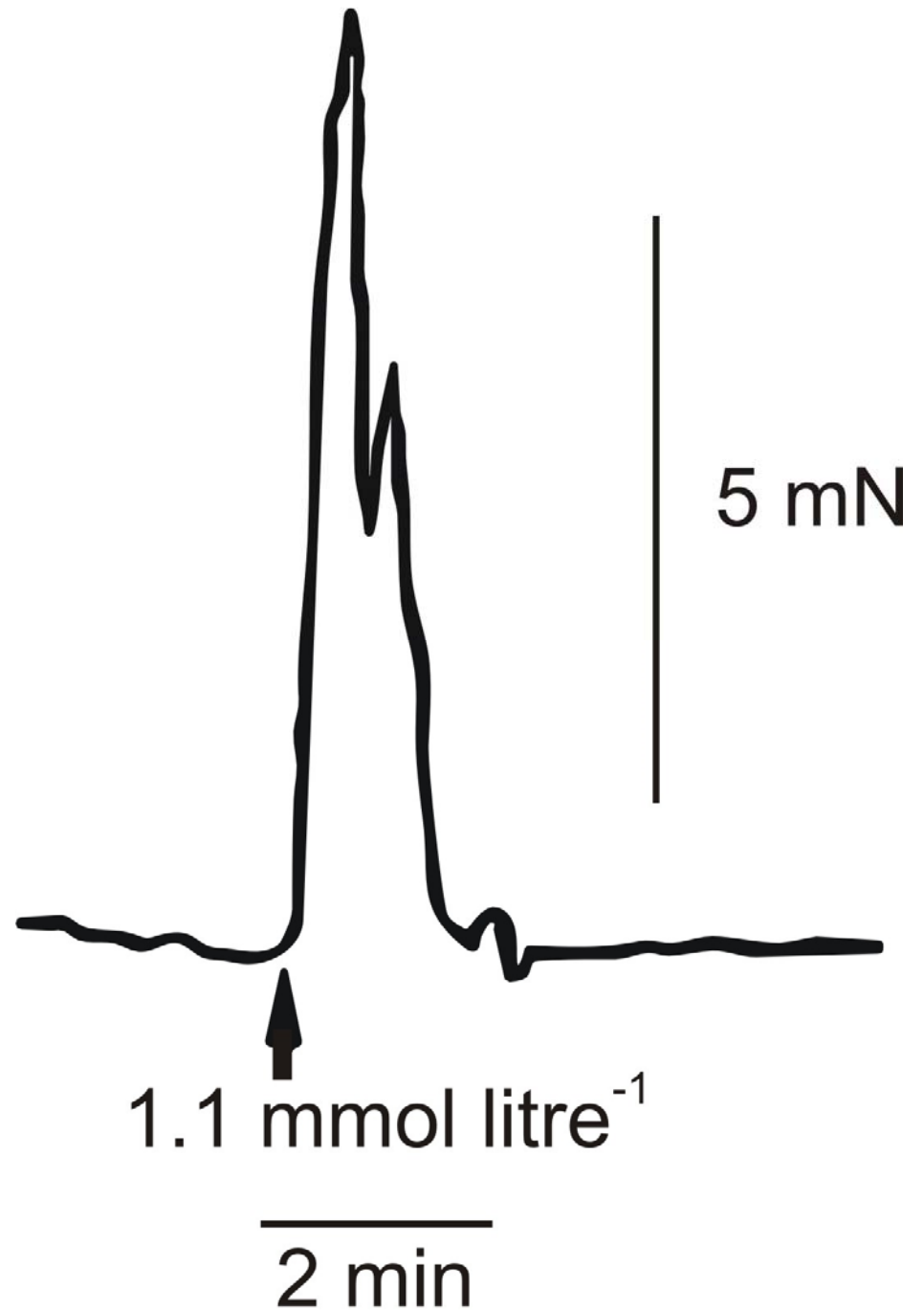
PP and general anaesthesia main problems

- 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)

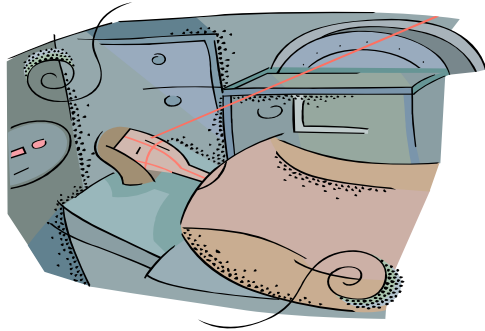
Complications

- 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 sarcoplasmic 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

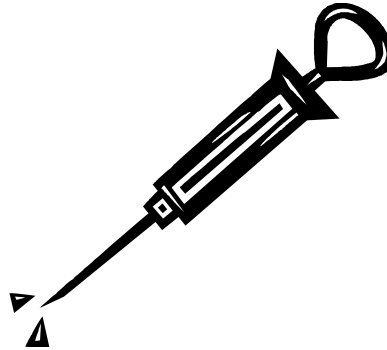
K kept in the high normal range

No pure NaCl or glucose infusions

Operation-induced stress to be reduced by Beta-Blocker

No Succinylcholine as relaxant (like for MH individuals)

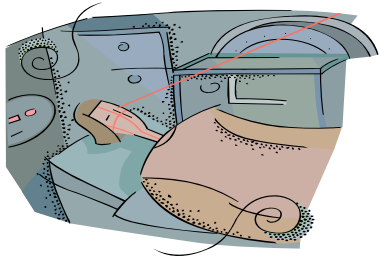
Local anesthesia and HypoPP



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 O₂, 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)