Use of Remifentanil in Pompe Disease: A Different Approach in a Changing Disease Process

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Introduction
- Pompe Disease is a deficiency of the lysosomal enzyme acid-glucosidase causing hypertrophic cardiomyopathy (HOCM) and muscle weakness.
- With early diagnosis and initiation of enzyme replacement therapy (ERT), patients have improved clinical outcomes, longer survival, and an increased need for surgical procedures requiring anesthesia.
- Techniques involving propofol, inhalational anesthetics, and ketamine have been described.
- We describe the use of remifentanil as an adjunct for anesthesia maintenance in an elective procedure for a child with Pompe Disease on ERT since infancy.

Case
An 11-year-old, 45.7kg, female with infantile-onset Pompe Disease is scheduled to have direct laryngoscopy and bronchoscopy to evaluate her existing tracheostomy, followed by replacement of a dislodged subcutaneous port.

Preoperative Assessment
- Patient at baseline respiratory status; on home ventilator FiO\textsubscript{2} 21%.
- Her most recent echocardiogram shows normal left ventricular (LV) size, end diastolic volume, and LV function without evidence of LV outflow tract obstruction. LV Mass Index 61.77 g/m\textsuperscript{2} (Z score -0.97).
- EKG showed sinus rhythm; no history of arrhythmias elicited.
- Review of prior anesthetic records.

Management
- Midazolam for preoperative anxiolysis.
- Induced with sevoflurane (etSev 2.3%) in a 50/50 O\textsubscript{2}/N\textsubscript{2}O mixture, a remifentanil infusion (0.1 mcg/kg/min), and fentanyl.
- During induction, patient noted to develop ST segment depression, which was treated with phenylephrine and intravenous fluids.
- Maintenance on sevoflurane (etSev 1.1-1.6%), increased remifentanil dose of 0.2 mcg/kg/min. Analgesia achieved with local anesthetic infiltration and acetaminophen.

Discussion and Conclusions
- This case illustrates the use of remifentanil supplementing a sevoflurane anesthetic in an infantile-onset Pompe Disease child who has been treated with disease-modifying ERT.
- Given her history of delayed emergence from anesthesia and her risk for hypertrophic cardiomyopathy, remifentanil was chosen.
- Despite the absence of cardiomyopathy on echocardiogram, the patient did not tolerate tachycardia and decreased coronary perfusion.
- An opioid-based technique was ideal for induction and maintenance of anesthesia, and remifentanil, combined with low dose sevoflurane, provided suitable conditions for a short, elective procedure.

Hemodynamic Goals for Pompe Disease Patients

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<tr>
<th>Hemodynamic Goal</th>
<th>Potential Agents to Use</th>
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<tbody>
<tr>
<td>Avoid myocardial ischemia (maintain coronary perfusion pressure)</td>
<td>Judicious use of propofol and inhalational anesthetics that ( \triangleright SVR, \triangleright DBP ), direct myocardial depressants</td>
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<td>Avoid decreased cardiac output</td>
<td>Adequate hydration (consider pre-op hydration) for adequate preload</td>
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<td>Consider ketamine IV which maintains SVR and contractility</td>
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<td>Balanced anesthetic with opioids to decrease dosages of maintenance agents</td>
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Preoperative Assessment
Review echocardiogram, assess for HOCM
EKG- assess for arrhythmia
If HOCM severe, consider preoperative IV hydration when NPO

Intraoperative management
Avoid high doses of agents that decrease SVR and contractility
Balanced anesthetic; consider short-acting opioids, like remifentanil
Judicious use of neuromuscular blockade
Regional anesthesia when feasible
Phenylephrine bolus available to increase diastolic blood pressure

Post-operative management
Routine management; discharge when patient at baseline respiratory status

REFERENCES