Anesthesia Recommendations for Patients Suffering from Prader-Willi Syndrome

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1. General considerations

- Rare genetic disorder (1:10.000 – 1:30.000 nn)\(^1\)
- Hipotalamic-pituitary disfunction
- The most frequent cause for “syndromic” obesity\(^1\)
- Newborns with marked hypotonia, of central origin
- Genitals with hypoplasia- cryptorchism
- Particular phenotype:
  - Narrow forehead
  - Almond-shaped eyes
  - **Sluggish saliva**
  - Turned mouth
  - Thin upper lip
Organ system involvement with PWS

### Airway
- Difficult tracheal intubation
- Small glottic opening
- Limited neck mobility
- Macroglossia

### Respiratory
- Hypotonia
- Poor cough – ineffective clearance
- OSA
- Intraoperative bronchospasm
- Restrictive lung disease

### Cardiovascular
- Ventricular arrhythmia
- Hypertension
- Cor pulmonale

### CNS
- Temperature instability
- Defective central control of ventilation
- Mental retardation
- Aggressive behavior

### Miscellaneous
- Obesity
- Glucose intolerance
- Kyphoscoliosis
- Poor dentition
- Viscous saliva and airway secretions
Ghrelin excess → Hyperphagia → Morbid obesity

RESP
- Hypoventilation
- ↓ SaO₂%
- Obstructive sleep apnea (OSA)

METAB
- Diabetes mellitus
- Debut at 20 years old
Growth hormone deficit
- Short stature
- Hypogonadism

Decreased bone mineral density

Cognitive impairment
- Learning disabilities
- Behavioral troubles

Impaired speech and language development
2. Typical surgery in the case of PWS patients

- Orthopedic surgery:
  - Spinal surgery – scoliosis
- Cleft lip, palatal repair
- Dental treatment due to tooth decay
- Orhidopexy - cryptorchism
3. Anesthesia – Particularities

- Morbid obesity
- Obstructive sleep apnea (OSA)
- Difficult peripheral intravenous cannulation
- Difficult airway management
- Risk for perioperative respiratory failure
- Primary myocardial involvement
- Aggressive and violent behavior
- Convulsions
- Disturbances in thermoregulation
- Glucose intolerance
4. Management in General Anesthesia

Additional preoperative diagnostic procedures

- BMI, neck circumference
- Dental issues
- Evaluation of current respiratory status
  - Polysomnography – identification of patients with severe OSA
  - PaO2%, PaCO2%, oxygen saturation
  - !!! Children with PWS frequently suffer from restrictive lung disease (due to hypotonia, obesity, and kyphoscoliosis)
- 12 lead ECG ± echocardiography
- EEG
- Test for CAI (Central Adrenal Insufficiency)
- Thyroid hormone levels – possible hypothiroidism
Obstructive Sleep Apnea (OSA)

- Polysomnography
- **STOP-Bang Questionnaire** (loud Snoring, Tiredness, Observed apnea, high blood Pressure – Body mass index, age, neck circumference, gender)
- Patients with OSA lose airway stability upon induction of GA → **ventilation may be difficult or impossible**
- After extubation – use non-invasive CPAP in order to stabilize the airway
Aspiration risk

- Hypotonia – unable to cough effectively after use of breathing tube
- Decreased motility of GI tract
- Physiological setpoint for vomiting is abnormal
- Obese → Higher incidence for hiatal hernia
- Low esophageal sphincter tone
Lower the aspiration risk

- Pre-anesthetic fasting
- Peripheral iv cannula
- Routine monitoring
- Drugs that reduce gastric volume and lower the acidity of gastric contents
- Premedication
- Rapid sequence induction
Pre-anesthetic fasting

**Table 2** Royal College of Nursing recommendations for preoperative fasting (see web link). Grades of recommendations: A = at least one meta-analysis, systematic review or randomized controlled trial; B = body of evidence applicable to the target population, demonstrating consistency of results D = non-analytic evidence (case reports, case series), expert opinion/consensus

<table>
<thead>
<tr>
<th>Grade</th>
<th>Recommendation</th>
</tr>
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<tbody>
<tr>
<td>A</td>
<td>Water and other clear fluids (through which newsprint can be read), clear tea and black coffee up to 2 h before induction of anaesthesia for elective surgery are safe and improve patient well-being</td>
</tr>
<tr>
<td>B</td>
<td>Tea and coffee with milk are acceptable up to 6 h before induction of anaesthesia</td>
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<tr>
<td>D</td>
<td>Minimum preoperative fasting time of 6 h recommended for food (solids and milk)</td>
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<tr>
<td>D</td>
<td>Breast milk may be given up to 4 h before induction; formula or cows’ milk up to 6 h before induction</td>
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<tr>
<td>B</td>
<td>Chewing gum should not be permitted on the day of surgery</td>
</tr>
<tr>
<td>D</td>
<td>Sweets (including lollipops) are solid food. Minimum preoperative fasting time of 6 h recommended</td>
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Peripheral intravenous cannula

- May be **difficult** due to the adipose tissue
- Should at first be attempted on the dorsal side of the hand
- **Ultrasound guided** iv cannula might be required
- Upon receiving informed consent the patient/parents should be warned of the eventuality of an **arterial cannula**, **central venous catheter** or other **invasive** monitoring techniques
Routine monitoring

- **Hemodynamic monitoring** - BP cuffs should be of appropriate size
- **Ventilation monitoring** – capnography (EtCO₂)
- **Fasting glucose** – at risk for perioperative hypoglycemia – even in the absence of diabetes (lipolysis and neoglucogenesis abnormalities)
- **Body temperature** – thermoregulation
- **Monitoring of the neuromuscular blockade**
  - NMT – Neuromuscular transmission
  - Entropy
- **Further monitoring in ICU** – at risk for: desaturation, hypercapnia, intermittent bronchospasm
Routine monitoring - Entropy

- Monitoring the state of the brain by data acquisition of EEG signals
- Monitoring the effects of certain anesthetic agents
- Help the user to titrate anesthetic drugs
- Reduction of anesthetic use and faster emergence from anesthesia
- Monitoring hypnotic effect of certain anesthetic drugs on the brain
Routine monitoring - NMT

- the transfer of an impulse between a nerve and a muscle in the neuromuscular junction.
- NMT can be blocked by neuromuscular blocking agents
- used to safely time extubation and avoid the occurrence of residual paralysis
- provides quantitative, automatic measurements of muscle response to stimulus and consequentially, the level of block
Aims of pharmacological premedication:

- Anxiolysis, sedation, amnesia
- Hemodynamic stability
- Lower gastric secretion, increase gastric pH
- Lower doses of anesthetic medication
- Facilitation of anesthesia induction
Premedication

- **BENZODIAZEPINES - MIDAZOLAM**
  - Preferred because of short $T\frac{1}{2}$ time
  - Sedative and anxiolitic
  - Amnesia
  - Doses: 0.07 mg/kg
  - !!! May lead to respiratory depression, airway collapse, respiratory failure, hypoxemia

- **OPIOIDS – FENTANYL**
  - !!! Associated with risk of respiratory distress
  - Doses are based on LBW (Lean Body Weight) - tissue that is metabolically active (increased due to obesity)
  - $LBW = IBW + 0.3 \ (TBW-IBW)$

!!! Prolonged and exaggerated response to sedatives and/or analgesic drugs is possible
Premedication

Drugs that lower gastric volume and gastric acidity

• **RANITIDINE**
  – H2 receptor blockade
  – Decreases gastric secretion as a response to histamine release, acetylcholine, and gastrin
  – Dose: 150 mg i.v., 1 h before surgery

• **METOCLOPRAMIDE**
  – Dopamine antagonist
  – Prokinetic
  – Pyloric sphincter relaxation – reduces gastric volume
  – Increases IES tone
  – Dose: 10 mg i.v., 1 h before surgery

Management of CAI

• **Hydrocortisone** - 100mg - 500 mg in adults; 0.56 si 4 mg/kg
Rapid sequence induction (RSI)

- Patient monitoring
- Preoxygenation – minimum 5 min.
- Sellick maneuver – continuous cricoid pressure starting when substances are administered and up until the airway is secured
- Anesthetics will be given fast, iv bolus
- Induction with Midazolam, Fentanyl, Propofol (2mg/kgc), and a neuromuscular blocking agent - Succinylcholine
- The patient will not be ventilated with facial mask
Rapid sequence induction (RSI)

Rapid Sequence Induction Checklist

**PATIENT**
- Pre-oxygenate
  - 3min >15L/min O₂
  - NIV if obese/OSA or SpO₂ <95% despite O₂
- Position
  - ‘ear to sternal notch – ‘RAMP’ if obese
- IF DIFFICULT AIRWAY ANTICIPATED CALL ANAESTHETIST IN CHARGE (ext: ___)
  - upper airway obstruction/trauma
  - morbidly obese/OSA
  - c-spine immobilisation

**EQUIPMENT**
- "SOAPME"
  - Suction
  - Oxygen
  - Bag-Valve Mask
  - Airway equipment
  - 2 laryngoscopes
  - 2 ETTs
  - bougie +/- stylet
  - Pharmacological agents
  - pretreatment
  - induction agent
  - paralytic agent
  - ongoing anaesthesia
  - fluids
  - vasoconstrictors
  - Monitoring Equipment
  - ETCO₂
  - SpO₂
  - ECG monitoring
  - NIBP

**TEAM**
- Team leader
  - Consultant if available
- Airway doc
  - Must have anaesthetic experience
- Airway nurse
- Drugs
  - JMO/nurse
- Scribe & timer
  - Nurse
  - Cricoid pressure
  - Optional JMO/nurse
  - IF DIFFICULT AIRWAY ANTICIPATED CALL ANAESTHETIST IN CHARGE (ext: ___)

**HAVE A PLAN**
- If you fail
  - See default strategy for failed RSI algorithm and let your team know if you are doing something different
- If you succeed
  - See Oxylog 3000 plus ventilator guideline

https://lifeinthefastlane.com/ccc/difficult-airway-algorithms/
**Neuromuscular blockade**

- During RSI – use short-acting depolarizing blocking agents – **Succinylcholine** (Lystenon)

- Use of non-depolarizing blocking agents for muscle relaxation should be restricted - potential long-lasting neuromuscular blockade

- Safe use for: **Rocuronium** – rapid reverse possible with Sugammadex (Ciclodextrina)

- Neuromuscular monitoring - residual blockade should be antagonized with **Neostigmine**
Airway management

- Difficult airway because of:
  - Thick saliva – complicates extubation
  - Facial dimorphism
  - Micrognatia
  - Palate anomalies
  - Impaired mobility of cervical spine and temporomandibular joint

- 13-25% - difficult intubation
- 2/3 difficult ventilation\(^2\)
- Anti-Trendelenburg position

!!! The appropriate equipment to deal with the "cannot intubate/cannot ventilate" scenario should be available
↑ risk for difficult airway

Fast desaturation

Preoxygenation

2-5 minutes on facial mask

- Positive pressure – CPAP – 8 cm H₂O
- FiO₂ 0.9-1
- Fresh gas 10 l/min

- Increases oxygen reserve
- Decreases severity of desaturation upon induction
- Reduces atelectasis formation
Airway management

Techniques for difficult intubation

• Alternative laryngoscope blades
• Awake intubation - video laryngoscope/fiberoptic scope
• Blind intubation (oral or nasal)
• Fiberoptic intubation
• Intubating stylet (Cook catheter or bougie)
• Invasive airway access
Airway management

**DEFAULT STRATEGY FOR FAILED RSI IN ADULTS**

**Plan A:**
- Initial tracheal intubation plan
  - Direct laryngoscopy
    - Maximum 2 attempts in 2 mins
    - Re-oxygenate if SpO₂ < 90% with 2 person BVM + OPA + NPA
    - Call anaesthetics if Plan A fails (ext: 3186)
  - RSI checklist
    - Pre-oxygenate
    - Position: ‘ear to sternal notch’ - ‘RAMP’ if obese
    - Paralysis & sedation for all
    - Cricoid pressure for all initially but release if poor view and apply External Laryngeal Manipulation
    - Bougie for all

**Plan B:**
- Secondary tracheal intubation plan
  - Video laryngoscopy
    - As difficult airway, maximise laryngeal view by avoiding cricoid pressure and by using External Laryngeal Manipulation

**Plan C:**
- Maintenance of oxygenation/ventilation
  - LMA
    - Avoid cricoid pressure
    - Improved oxygenation

**Plan D:**
- Rescue techniques for “can’t intubate can’t ventilate” situation
  - Scalpel/ finger/ tube cricothyroidotomy

Contact anaesthetics (ext: _____) for fibroptic intubation

[https://lifeinthefastlane.com/ccc/difficult-airway-algorithms/](https://lifeinthefastlane.com/ccc/difficult-airway-algorithms/)
Maintenance of anesthesia

- **Propofol** – TCI
- Isoflurane
- Sevoflurane

- **Ketamine**
  - also able to provide sedation and analgesia
  - Limited effects on respiratory function
  - Should be administered with propofol or a benzodiazepine to limit potential emergence phenomena
5. Regional anesthesia in Prader-Willi syndrome

- Eliminates perioperative risks of GA
- Eliminates the need for GA
- Provides
  - Intraoperative anesthetic care
  - Postoperative analgesia → limiting the need for opioids
- Peripheral nerve blockade
- Neuraxial techniques (epidural or spinal anesthesia)
5. Regional anesthesia in Prader-Willi syndrome

- Landmarks for regional anesthesia may be obscured due to morbid obesity
- Use ultrasound to facilitate placement of the block
- Sedation needed for
  - Facilitating placement of the block
  - Limit incidence of complications
- Use *routine monitoring*
- Be ready to intubate if loss of airway
References

3. Jon Roberts, Mary Cataletto et al, *Special Anesthesia Concerns for Patients with Prader-Willi Syndrome: The Winthrop University Hospital Center Experience*, The Gathered View, y by the Prader-Willi Syndrome Association (USA), ISSN 1077-9965
8. https://lifeinthefastlane.com
Thank you!