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Case Report

High flow nasal oxygenation and videolaryngoscopy - the two saviours in a case of Prader-Willi syndrome: A case report

Aparna Singh[1], **Prachi Gaba**[1]*, **Kush Sharma**[2], **Lalit Gupta**[1]

[1] *Dept. of Anaesthesiology and Critical Care, Maulana Azad Medical College and Associated Hospital, Delhi, India*
[2] *Rajiv Gandhi Super Speciality Hospital, Delhi, India*

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ABSTRACT

Prader-Willi syndrome (PWS) is a rare genetic disorder characterized by hypothalamic-pituitary abnormalities, hypotonia, hyperphagia, and a high risk of morbid obesity, which complicates both general and regional anesthesia. Patients with PWS face additional challenges, including disturbances in thermoregulation, arrhythmias, cor pulmonale, and convulsions. These factors make anesthetic management particularly challenging, necessitating careful planning and monitoring.

We report the anesthetic management of a 7-month-old infant with genetically confirmed PWS who underwent bilateral orchidopexy under general anesthesia. The patient's preoperative evaluation revealed significant hypotonia and a predisposition to respiratory complications. During the procedure, advanced airway management techniques, including the use of C-MAC and high-flow nasal oxygenation (HFNO), were employed to alleviate the risk of difficult airway and respiratory failure. The anesthesia team meticulously monitored the patient's vital signs and thermoregulation throughout the surgery to prevent arrhythmias and other complications associated with PWS.

The successful anesthetic management of a child with Prader-Willi syndrome requires a multidisciplinary approach, thorough preoperative planning, meticulous intraoperative monitoring, and vigilant postoperative management to minimize the risk of complications in this vulnerable patient population.

1. Introduction

Prader-Willi syndrome (PWS) is a rare neurodevelopmental disorder occurring in approximately 1 in 15,000 to 30,000 live births. It results from abnormalities on chromosome 15q11.2-q13, typically due to paternal deletion or maternal uniparental disomy.[1] PWS is characterized by a biphasic presentation: an infantile phase of hypotonia and poor feeding, followed by childhood hyperphagia leading to obesity. While early phase is characterized by delayed milestones, the later phase is characterized by hyperphagia leading to childhood obesity along with behavioural disturbances, convulsions and arrhythmias.[1–3]

The genetic basis of PWS has significant implications for anesthetic management. Hypothalamic dysfunction leads to various endocrine abnormalities, while hypotonia and obesity contribute to respiratory complications. These factors, combined with potential difficult airways, difficult intravenous cannulation, and postoperative respiratory failure leading to ventilator dependency pose substantial challenges for anesthesiologists.

We present the case of a 7-month-old infant with PWS undergoing bilateral orchidopexy, highlighting the use of video laryngoscopy for airway management and high-flow nasal oxygenation (HFNO) for postoperative respiratory support.

* *Corresponding author*.
E-mail address: aparna.singh.raghuvanshi02@gmail.com (P. Gaba).

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2. Case Presentation

2.1. Preoperative assessment

A 7-month-old male infant with genetically confirmed PWS was scheduled for laparoscopic orchidopexy. Born prematurely at 34 weeks gestation, the patient had a history of neonatal respiratory distress requiring oxygen support under oxygen hood for 24-48 hours in neonatal intensive care unit. Developmental milestones were delayed, and poor sucking reflex necessitated spoon feeding.

Physical examination revealed an obese infant weighing 9.65 kg with inspiratory stridor and intercostal retractions, improving in the prone position. Airway assessment showed a short, thick neck with limited mobility and macroglossia. Cardiovascular evaluation, including ECG and echocardiography, was unremarkable.

2.2. Anesthetic management

In the operating room, standard ASA monitoring was applied. A difficult airway cart was prepared, including various laryngoscope blades, intubating stylets, supraglottic airways, and equipment for emergency surgical airway.

Anesthesia was induced with sevoflurane 4-8% in oxygen while maintaining spontaneous respiration. After securing intravenous access, fentanyl (1 μg/kg) was administered. Neuromuscular monitoring was established, and atracurium (0.5 mg/kg) was given to facilitate intubation.

Laryngoscopy was performed using a C-MAC video laryngoscope with a paediatric D-blade, chosen for its potential advantages in difficult paediatric airways. A grade 2 view (POGO score 20%) was achieved, and a 3.5 mm uncuffed endotracheal tube was successfully placed. During laparoscopy, end-tidal CO_2 increased to 60 mmHg, necessitating adjustments in ventilation strategy to maintain end-tidal CO_2 between 40-45 mmHg. Temperature was closely monitored throughout the procedure.

2.3. Postoperative care

Following reversal of neuromuscular blockade, the patient was extubated when showing adequate spontaneous respiratory efforts. Post-extubation stridor and tachypnoea developed, initially managed by placing the infant in a prone position, which relieved the obstruction and improved his respiratory pattern to some extent. Over a period of 30 minutes, baby's condition improved further and he was maintaining 99% saturation on oxygen but with the use of accessory muscles. High Flow Nasal Oxygenation (HFNO) at 16L/min, 0.40 $fiO_2$was started and patient was shifted to PICU. He was weaned off of HFNO over 48 hours and discharged from the hospital after 5 days.

3. Discussion

Prader-Willi syndrome is linked with absence of paternal genes on chromosome 15q11-13 and is characterized by hypotonia, obesity, hypogonadism and developmental delay. It was first described by Prader, Labhart, and Willi in 1956.[4] Classically it has a biphasic presentation, consisting of an early infantile hypotonic phase and an obese childhood phase. Both general and regional anaesthesia are known to be quite intimidating in patients with PWS. Difficult bag and mask ventilation as well as difficult intubation should be anticipated while instituting general anaesthesia. Landmarks for regional anaesthesia may be obscured due to morbid obesity. Other anaesthesia considerations are difficult intravenous cannulation, thermoregulation, perioperative risk of aspiration and metabolic disturbances. Presence of Obstructive Sleep Apnea (OSA) can make the matters worse, and is one of the main reasons behind prolonged hospital stay in PWS patients who received general anaesthesia.[5]

Tailoring the anaesthesia specific to the needs of these patients like limiting the use of opioids in intraoperative and postoperative period and avoiding neuromuscular agents, reduces the severity of OSA after extubation. The decision to use the C-MAC video laryngoscope with a D-blade was based on recent literature suggesting its efficacy in paediatric difficult airways. While the usefulness of the D-Blade has been reported for intubation of adults since 2011,[6] two infant manikin studies of difficult airway situations, demonstrated a better glottic view and faster intubation while using C-MAC D-blade as compared to conventional direct laryngoscopy.[7,8] The higher angulation (40 degrees) and distal optical camera near the blade tip of the D-blade provide an improved glottic view without requiring alignment of the oral, pharyngeal, and laryngeal axes. However, a stylet is often required for intubation due to the extreme curvature of the D-blade. This proved advantageous in our patient with limited neck mobility and macroglossia. Our alternative plan, should this have failed would have been LMA blockbuster insertion followed by fibreoptic guided intubation.

Postoperative hypoxia is a significant concern in PWS patients due to obesity, hypotonia, and potential kyphoscoliosis.[9] Deranged awake responses to changes in blood oxygen and CO_2 content in PWS patients is considered to be another cause of post operative hypoxia. Our use of HFNO in the immediate postoperative period addressed this risk effectively. HFNO provides several benefits, including upper airway dead space washout, provision of positive airway pressure, and improved oxygenation. The high gas flow leads to upper airways dead space washout creating an oxygen reservoir within the upper airways, leading to improvement in vital signs, blood gas values and reduced incidence of intubationsin patients with respiratory failure. Additionally, a flow-dependent effect of

continuous positive airway pressure has been documented in healthy subjectsand in patients.[10–12] While studies on HFNO in paediatric patients are limited, our experience suggests it can be a valuable tool in managing postoperative respiratory distress in PWS patients.

The main limitation of our approach was the necessity for general anesthesia with muscle relaxation due to the nature of the surgery. Regional anaesthesia techniques, although difficult due to masked anatomical landmarks are the preferred modality in PWS, since they eliminate the need for GA and hence reduce the perioperative risks of respiratory failure and aspiration due to restrictive lung disease, hypotonia and postoperative residual neuromuscular blockade.[13]

4. Conclusion

Successful anesthetic management of infants with PWS requires thorough preoperative evaluation, careful airway management, and vigilant postoperative respiratory monitoring. The use of video laryngoscopy for intubation and HFNO for postoperative support proved effective in our case. Future research should focus on developing standardized protocols for anesthetic management of PWS patients and further evaluating the efficacy of HFNO in this population.

5. Source of Funding

None.

6. Conflict of Interest

None.

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Author's biography

Aparna Singh, Senior Resident https://orcid.org/0000-0002-8907-7471

Prachi Gaba, Specialist https://orcid.org/0000-0002-7878-9879

Kush Sharma, Assistant Professor https://orcid.org/0000-0002-1078-1010

Lalit Gupta, Professor https://orcid.org/0000-0001-7291-5961

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