Case Report

A Case Report of Facing Challenges During Anesthetic Management in a Patient with Prader–Willi Syndrome Undergoing Laparoscopic Surgery

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ABSTRACT

Background: Prader-Willi syndrome (PWS) is a heterogeneous genetic disorder caused by an anomaly on the chromosome 15. This condition typically results in low muscle tone, reduced height, cognitive impairments, and persistent feelings of hunger, which can lead to severe obesity. Additional common challenges associated with PWS include disturbances in thermoregulation, arrhythmia, diabetes mellitus, and seizures. These multisystemic issues create unique challenges in anesthetic management, making thorough perioperative planning essential to minimize potential complications.

Case Presentation: We present the case of a 19-year-old male with PWS who underwent laparoscopic hernioplasty and cholecystectomy under general anesthesia. The perioperative course was largely uneventful, with successful weaning from mechanical ventilation. The only notable occurrence was transient, intermittent hypertension during pneumoperitoneum, which was effectively managed without complications.

Conclusion: Careful perioperative planning and multidisciplinary management enable safe anesthesia and favorable surgical outcomes in patients with Prader–Willi syndrome.

Keywords: *Prader-Willi syndrome, Anesthetic Management, Opioid-Sparing Analgesia, Difficult Airway, Laparoscopic Surgery, Pneumoperitoneum.*

INTRODUCTION

Prader-Willi syndrome (PWS), originally described in 1956, is a genomic imprinting disorder affecting chromosome 15q11-13[1]. It was first described in 1887 by John Langdon Down in a patient with "polysarcia". In 1956, Prader, Labhart and Willi described nine other case, the classic clinical phenotype, from whom the syndrome derives its name. and gave the syndrome its name[2]. The multisystem nature of PWS characterized by muscular hypotonia, central nervous system abnormalities, behavior problems, obesity, hypogonadism and skeletal abnormalities presents a unique set of challenges for the anesthesiologist. Decreased pulmonary reserve secondary to chest wall deformity (i.e., scoliosis), hypotonia and obesity may complicate ventilator management during and following anaesthesia[3]. Major challenges faced by an anesthesiologist in the perioperative management of PWS is summarized in Table-1.

The comprehensive anesthetic approach should include all the associated comorbidities[4]. Special attention to the potential difficulties in the difficult airway management, behavioral problems, OSA, hemodynamic instability, hypotonia should be addressed by the anesthesiologist. This case report outlines the challenges faced during anesthetic management for a PWS patient scheduled for laparoscopic hernioplasty and cholecystectomy.

CASE PRESENTATION

A 19-year-old male with diagnosed Prader-Willi syndrome (PWS) was scheduled for laparoscopic mesh hernioplasty and cholecystectomy at Evercare Hospital Dhaka. His presentation was notable for multiple PWS-associated comorbidities: morbid obesity, short stature, diabetes mellitus, obstructive sleep apnea, hypogonadotropic hypogonadism, behavioral disability, osteoporosis and myopia.

Table 1: Anesthetic concerns in PWS

Catogory	Key Concerns
Airway Challenges	- Obesity,micrognathia, - high-arched palate - Thick saliva and hypotonia
Respiratory Risks	- Increased risk of airway obstruction - Obstructive sleep apnea (OSA) - Hypoventilation due to hypotonia - Restrictive lung pattern - High risk of postoperative failure
Metabolic Issues	- Obesity, insulin resistance - Type 2 diabetes mellitus - Risk of perioperative hypoglycemia or hyper glycemia
Thermoregulatory Defects	- Hypothalamic dysfunction - Impaired temperature regulation
Cardiovascular Risks	- Hypertension - Arrhythmias - Pulmonary hypertension
Endocrine Dysfunction	- Growth hormone deficiency - Hypothyroidism - Adrenal insufficiency - Impaired stress response to anesthesia
Behavioral & Cognitive Issues	- Anxiety, aggression, poor cooperation - Challenges with premedication and induction
Pharmacologic Sensitivities	- Altered drug metabolism - Increased sensitivity to sedatives and muscle relaxants

A thorough history was obtained during the pre-anesthesia check-up indicated that the patient is the youngest of three siblings, born to parents who are related by consanguineous marriages. He was delivered via caesarean section at term, experiencing delayed crying immediately after birth, with a birth weight of 2.2 kg. Developmental milestones were delayed, with hyperphagia onset at six months of age. He was diagnosed with diabetes at the age of nine and is currently managed with a regimen of mixed insulin and oral hypoglycemics. Additionally, he received hormonal treatment to address his growth and hyperphagia. He required admission for diabetic ketoacidosis at the age of fifteen and was subsequently diagnosed with PWS through genetic testing at Bangladesh Medical University.

During the preanesthetic checkup, Physical examination revealed classic dysmorphic features of PWS, including a narrow forehead, almond-shaped eyes, and small hands and feet with diminished gripping strength (Figure-1). The patient was hemodynamically stable, weighing 69 kg and standing 130 cm tall, which resulted in a BMI of 40.8. No abnormalities were noted during other general or systemic examinations. With a MET of over 4, the patient experiences difficulties in mobility due to obesity. An airway examination revealed Mallampati III classification, thick secretions, irregular dentition, and a high neck circumference of 18 inches, indicating potential challenges for airway management.

Preoperative investigations were significant for poorly controlled diabetes (random blood glucose 14.6 mmol/L, HbA1c 9.6%), sinus tachycardia on ECG, and pulmonary function tests indicative of a combined restrictive-obstructive pattern. Fiberoptic laryngoscopy revealed an enfolded epiglottis. Echocardiogram, thyroid, and liver function tests were within normal limits.

Given the compromised respiratory function and difficult airway, a comprehensive plan for continued postoperative ventilation was discussed and explained to the patient's attendant. The patient was also advised on managing blood sugar levels and engaging in breathing exercises.

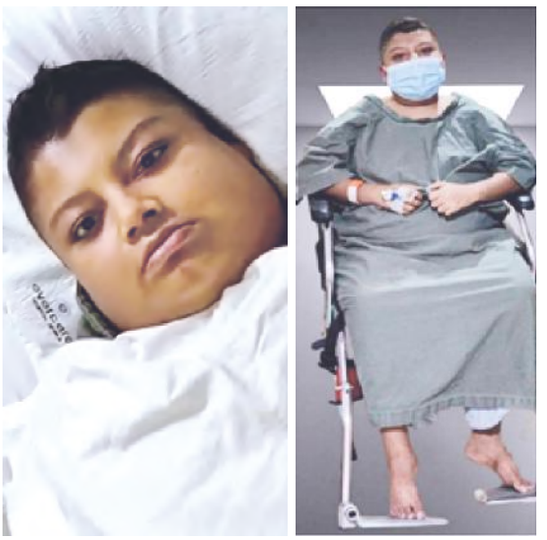

Figure 1: Provided facial features of PWS (Produced with patient's parent´s permission)

Notably, the initial surgery was postponed due to preoperative agitation and anger tantrums prior to transfer from the ward to the operating theatre. Upon readmission, this was managed with overnight oral midazolam to ensure proper cooperation and maintain fasting. Peripheral venous access was established with relative ease. In advance of the procedure, difficult airway management equipment and a fiberoptic bronchoscope were readily available. Continuous monitoring included standard parameters, invasive blood pressure, train-of-four (TOF), Bispectral Index (BIS) and core temperature monitoring.

Anesthesia was successfully induced using Midazolam, along with a target-controlled infusion (TCI) of Remifentanil and Propofol, with cisatracurium employed for neuromuscular blockade. Endotracheal intubation was successfully achieved with a video laryngoscope (Cormack-Lehane grade 2). Patient warmer with infusion of warm IV fluid aided to maintain the core temperature at the range of 36-38°C. A bilateral ultrasound-guided quadratus lumborum block was performed for multimodal analgesia (QL block is a fascial plane block which allows local anesthetic spread to thoracolumbar nerves T7 -L2 which supply sensation to the abdominal wall and sympathetic trunk contributing visceral pain relief) followed by placement of A 16F nasogastric tube and urinary catheter . After the establishment of pneumoperitoneum, volume control ventilation was transitioned to pressure support mode due to elevated airway pressures. Incremental doses of Cisatracurium were titrated to maintain a TOF of 0–1, with the return to TOF of 1 occurring at the anticipated interval of 30 minutes.
During the intraoperative period, the patient experienced hypertension, which was effectively managed using intravenous labetalol and a glyceryl trinitrate (GTN) infusion. Anesthesia maintenance involved Target controlled infusions of Propofol and Remifentanil over the duration of the surgery, which lasted 3.5 hours. Both preinduction and post-procedure arterial blood gas analyses were within normal limits, showing no significant differences.

Postoperatively, the patient was transferred to the surgical intensive care unit with continued mechanical ventilation. Gradual weaning commenced on postoperative day 1, transitioning from volume control to synchronized intermittent mandatory ventilation (SIMV) mode, with successful extubation achieved thereafter, leading to a transfer to the ward. Postoperative complications included new-onset hypertension, requiring initiation of antihypertensive medication, and copious thick secretions managed with frequent suctioning, which resolved by postoperative day 3. Blood glucose levels were effectively managed with previously established medications. Pain management included intramuscular Pethidine 75 mg administered on demand, complemented by regular intravenous Paracetamol every eight hours. The patient was discharged on the fourth postoperative day, exhibiting stable vital signs.

DISCUSSION

The perioperative management of patients with Prader-Willi syndrome presents a significant challenge to the anesthesiologist, necessitating a meticulous, multidisciplinary approach tailored to the syndrome's unique pathophysiology. Our case illustrates the successful application of such a strategy for a major laparoscopic procedure, highlighting key considerations for airway management, ventilation, analgesia, and postoperative care

Prader-Willi syndrome (PWS) is a multisystem genetic disorder caused by a lack of expression of paternal alleles in the PWS region of chromosome 15q11–13.5 This can occur through multiple genetic mechanisms including paternal deletion, uniparental disomy, mutation of the imprinting control center, or parental chromosomal translocation[6,7]. Diagnosis of PWS should be based on clinical criteria (Holm's criteria of 1993, revised in 2001) with confirmation by genetic analysis. It is described as a 2-stage disorder with an infantile hypotonic phase followed by a childhood obese phase[8]. It has an estimated prevalence of 1 in 10,000–30,000 live births.[9,10] The annual mortality rate was 3% for all ages, but the rate increases to 7% over the age of 30[11].

In individuals with PWS, morphological and pathophysiological alterations may lead to considerable respiratory complications, including a higher risk of perioperative respiratory complications i.e. regurgitation and aspiration, and the tendency towards post-surgical hypoventilation. These patients frequently have obstructive sleep apnea and restrictive patterns on pulmonary function tests, which leads to difficulty in respiratory management perioperatively[12], these factors predispose patients to rapid desaturation, difficult ventilation, and contributing to a significantly elevated risk of sudden death from respiratory causes. Our management anticipated these challenges. The predicted difficult airway (Mallampati III, high neck circumference, enfolded epiglottis) was secured using a video laryngoscope, minimizing trauma and attempts. Postoperatively, the planned transfer to the ICU for supervised, gradual ventilator weaning was a critical factor in avoiding respiratory failure, a strategy strongly recommended in the literature for patients with severe OSA[9]. Further difficulties may arise from mental retardation, anger tantrums, disturbed thermoregulation, and an unstable glucose metabolism[13]. These patients develop hypothalamic dysfunction that can lead to various endocrine changes. Familiarities with these issues can help to facilitate the anesthesia experience and prevent the number and severity of postoperative complications[14]. The summary of pathophysiology of PWS is depicted in Figure-2.

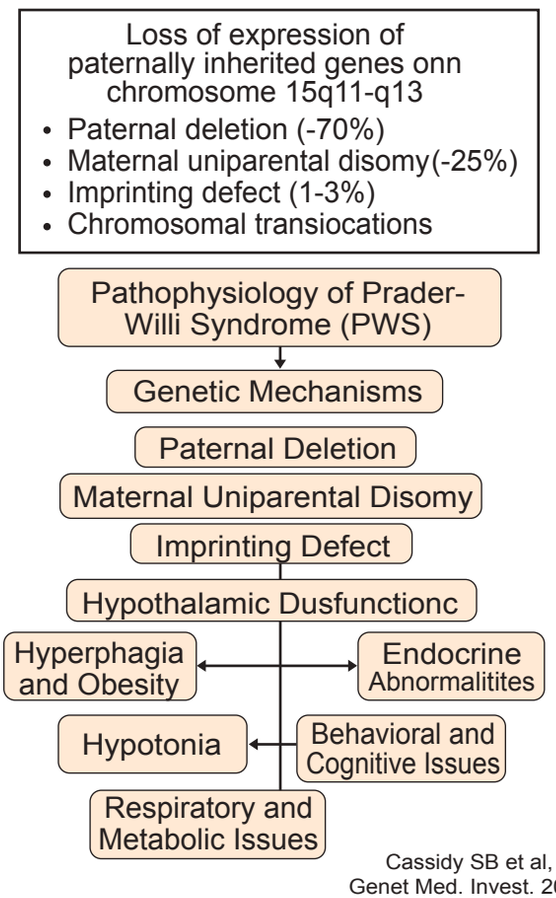

Figure 2: Genetic mechanisms and hypothalamic dysfunction in the pathophysiology of Prader–Willi Syndrome (PWS), leading to obesity, endocrine, cognitive, and metabolic complications (Cassidy SB et al., Genet Med. Invest. 2015)

There have been reports of delayed recovery in these patients[15]. The titration of propofol (2-4µg/ml) for induction was guided by the Bispectral Index (BIS) score. We employed a lower propofol dose based on lean body weight, utilizing the BIS value for a more precise prediction of loss of consciousness.

Analgesia was maintained by TCI using Remifentanil (4-8 nanogram/ml), QL block and IV Paracetamol. Avoidance of neuromuscular blockade as part of anesthesia management is known to reduce the risk of airway and respiratory problems in patients with PWS[16,17], this may not always be feasible, as demonstrated in this case. Anesthetic management using atracurium and rocuronium have been reported[18]. Following the creation of pneumoperitoneum, there was an increase in airway peak pressure. Excessive intra-abdominal pressure decreases the pulmonary volumes in morbidly obese patients under anesthesia.4 In this case, the elevated peak pressure was addressed by adjusting the ventilator settings to pressure support mode, maintaining a pressure of 20 cmH_2O to achieve the necessary tidal volume.

Patients with PWS may experience disordered thermoregulation due to hypothalamic dysfunction. However, in this case, the patient's body temperature was effectively regulated using active warming measures such as forced warming blanket. Cardiovascular complications can include arrhythmias, conduction abnormalities, premature ventricular

contractions, and obesity-related hypertension.[1] Our patient exhibited elevated blood pressure during pneumoperitoneum, which was managed with a combination of intravenous labetalol and glyceryl trinitrate infusion, guided by arterial blood pressure monitoring. Following the procedure, he was prescribed oral ramipril for a new diagnosis of hypertension. Pulmonary hypertension, right-heart failure, and edema may necessitate further evaluation in these patients[19].

Postoperative intensive care is highly recommended for patients with PWS who have severe OSA to mitigate the risk of sleep-related respiratory complications, even in the absence of narcotics or intermediate to long-acting muscle relaxants. In this case, successful extubating and the avoidance of pulmonary complications were achieved by transferring the patient to the intensive care unit (ICU) and facilitating a gradual weaning process.

Additionally, preoperative fasting may be challenging due to hyperphagia, necessitating close monitoring by caregivers. Maintaining stable blood glucose levels can also be problematic due to alterations in carbohydrate and lipid metabolism; in this instance, the patient demonstrated a random blood sugar level of less than 10 mmol/l during the perioperative period. Individuals with PWS may exhibit a different pain response compared to others, which can obscure the identification of other issues. Pain management for this patient was successfully accomplished using a quadratus lumborum block and intramuscular pethidine, with only a single dose of pethidine required.

The safe anesthetic management of patients with Prader-Willi syndrome hinges on a thorough understanding of its multisystem pathology, with key principles including safe anesthetic management of Prader-Willi syndrome requires anticipating a difficult airway with advanced equipment, using titrated low-dose anesthesia to address drug sensitivity, adopting a multimodal opioid-sparing plan, closely monitoring autonomic and temperature instability, and providing postoperative intensive care with ventilatory support when needed; with careful preparation and tailored management, complex surgeries can be performed safely with good outcomes. This case demonstrates that with meticulous preoperative planning, tailored intraoperative management, and structured postoperative care, patients with PWS can undergo complex surgeries safely and achieve good outcomes.

CONCLUSION

The anesthetic management of patients with Prader-Willi Syndrome (PWS) requires a thoughtful approach to address potential airway challenges, stringent control of ventilation during surgery, effective management of metabolic disturbances, and robust cardiocirculatory support. A thorough pre-operative workup by the anesthesiologist, endocrinologist, pulmonologist, and pediatrician for identification and optimization of co-morbidities would go a long way in planning for perioperative management. Consequently, a detailed preoperative evaluation, meticulous intraoperative preparation, and vigilant postoperative monitoring in an appropriate care setting are strongly recommended for PWS patients undergoing surgical procedures.

INFORMED CONSENT

Informed consent was taken.

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