



ORIGINAL RESEARCH PAPER

Anesthesiology

LAPAROSCOPIC BARIATRIC SURGERY IN PRADER WILLI SYNDROME: ANAESTHESIA CONCERNS

KEY WORDS: Prader-Willi syndrome (PWS), Laparoscopic bariatric surgery, Anaesthesia concerns.

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ABSTRACT

Prader-Willi syndrome (PWS) is a rare complex multisystem genetic disorder of chromosome 15 resulting in physical, mental, and behavioural problem, especially unrelenting feeling of hunger causing morbid obesity. Anaesthesia concerns of PWS include morbid obesity, the potential for difficulties with airway management, risk for peri-operative respiratory failure, abnormalities in the central control of ventilation and temperature, rare reports of primary myocardial involvement, aggressive and at times violent behaviour and glucose intolerance. We present here the anaesthetic management of a 14-year-old female patient with Prader-Willi Syndrome, associated with morbid obesity (BMI 47.39), mild mental retardation, hypothyroidism and obstructive sleep apnoea syndrome, who was scheduled to undergo laparoscopic bariatric surgery. Anaesthetic considerations are reviewed highlighting perioperative complications associated with this syndrome.

INTRODUCTION

Prader Willi Syndrome (PWS; JL Down 1887) is a rare complex multisystem genetic disorder. Some genes on chromosome 15 are deleted or unexpressed, resulting in physical, mental and behavioural problems, especially unrelenting feeling of hunger causing morbid obesity. In 1956, Prader, Labhart and Willi (SWISS) gave the syndrome its name. With a prevalence of approximately 1/25000, PWS is the most common genetic cause of life threatening morbid obesity in children. Anesthetic considerations are reviewed highlighting perioperative complications associated with this syndrome

CASE REPORT

14-year-old girl affected by PWS (based on clinical examination and referred genetic analysis) associated with morbid obesity (Wt-130 kg, BMI - 46.7 kg/m²), irritable behaviour, uncontrolled feeling of hunger & OSA was scheduled for elective laparoscopic bariatric surgery under general anaesthesia. No history of defective central control of ventilation, cardio circulatory abnormality, gastro-oesophageal reflux, reduced salivary flow, previous episodes of laryngo-spasm or broncho-spasm. Airway assesment to rule out the presence of micrognathia, mandibular hypoplasia, poor dentition, large tongue, palatal abnormalities, adenoid hypertrophy and limited neck mobility. Physical & systemic examination was within normal limit. Routine preoperative and biochemical investigations were normal. ECG, C-XRAY, ABG, PFT were normal.

ANAESTHETIC MANAGEMENT

PREOPERATIVELY:

- To optimize patient condition: physiotherapy, diet counselling, regular respiratory exercise, for 2 days.
- Proper written and verbal consent from patient and attendants.
- NBM 12 hours.
- oral anti-aspiration prophylaxis given.

INTRAOOPERATIVELY:

ECG, SpO₂, NIBP, Urine output, E.T.T. Cuff pressure were monitored. Premedication was done with Inj. Glycopyrrolate 0.5 mg iv and Inj. Fentanyl 100 mcg iv. After preoxygenation for 5 mins induction was carried out with Inj. i/v Propofol 1% 150 mg, I/V succinylcholine 150mg, IPPV was done for 1min. Oral tracheal intubation was done with 7 mm ETT (cuffed). ETT cuff pressure was observed at various steps of bariatric surgery & kept adjusted at about 28cm H₂O by calibrated manometer. Anaesthesia was maintained with Oxygen :Air (60: 40), Desflurane 3% , I/V atracurium 40 mg..10mg... Surgery lasted for approximately 60 mins. Inj Paracetamol 1 gm iv was given for pain management and Inj Dexamethasone 8mg was given. Neuromuscular blockade was reversed with inj. neostigmine 2.5 mg and inj glycopyrrolate 0.4 mg iv. Patient was extubated uneventfully (following command & good tidal volume / minute volume achieved) Spo₂ @ room air was 100% and then shifted to intensive care

POSTOPERATIVELY:

Patient was shifted to the special care unit with oxygen by mask in propped up position. Patient was conscious, alert, oriented. Patient's vital parameters were normal.

DISCUSSION

CHALLENGES	COMPLICATIONS	MANAGEMENT
PWS Defective central control of ventilation	• hypoxia	• intra-operatively strict control of ventilator modality.
Temprature regulatory abnormalities	hypothermia / hyperthermia	• warm air blanket. • warm fluid to prevent hypothermia.
Thick saliva	• airway obstruction, suction blockage	• avoid antisialagogue drugs.
Bronchospasm	• oxygen desaturation	• selection of sevoflurane for inhalational anaesthesia (less irritative, bronchodilator)
Metabolic disturbances	hypoglycaemia	• monitor blood sugar intra & peri operatively.
Aggressive personality	desaturation / apnoea	• Avoid postop sedation and benzodiazepine.

Food seeking behaviour	<ul style="list-style-type: none"> increase risk of aspiration 	<ul style="list-style-type: none"> fasting must be verified by caregiver / put RT to assure empty stomach.
high pain threshold	<ul style="list-style-type: none"> mask underlying problems 	<ul style="list-style-type: none"> other possible sign of underlying problems should be monitored
Hypotonia	<ul style="list-style-type: none"> ineffective cough & inability to clear the airway after extubation. 	<ul style="list-style-type: none"> judicious use of muscle relaxant proper suctioning
growth hormone deficiency	<ul style="list-style-type: none"> may have smaller airway – difficult mask ventilation difficult intubation 	<ul style="list-style-type: none"> difficult airway management- induction in head up position. mask ventilation by 4 handed technique. appropriate equipment to deal with the “cannot intubate/cannot ventilate” scenario
OBESITY: OSA	<ul style="list-style-type: none"> apnoea / respiratory obstruction (sensitive to narcotic and sedative). 	<ul style="list-style-type: none"> Short acting narcotics intraoperatively avoid tranquilizers postoperatively. Naso-pharyngeal Airway.
low respiratory reserve volume (decrease FRC,FVC,TLC)	<ul style="list-style-type: none"> respiratory insufficiency (difficulty in ventilation, faster desaturation) 	<ul style="list-style-type: none"> head up position proper oxygenation
increase intra-abdominal pressure /GERD	<ul style="list-style-type: none"> increase risk of aspiration 	<ul style="list-style-type: none"> put efficient suction device very close. pre-op fasting, aspiration prophylaxis.
SURGERY Alteration in E.T.T. cuff pressure at various steps of surgery	<ul style="list-style-type: none"> increase cuff pressure-cough,sore throat, hoarseness of voice. decrease cuff pressure- risk of aspiration 	<ul style="list-style-type: none"> monitor & keep adjusted by manometer
haemodynamic changes	<ul style="list-style-type: none"> haemodynamic instability 	<ul style="list-style-type: none"> monitoring proper fluid management cardio-circulatory support

CONCLUSION

Individuals with PWS can safely undergo anaesthesia. Anaesthetic management of these patients includes a careful approach to difficulties with airway, strict control of intraoperative ventilation, management of metabolic disturbances and cardio circulatory support.

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