Anaesthetic management of paediatric patient with Prader-Willi syndrome for bariatric surgery

Sir,

Prader –Willi syndrome (PWS) is characterized by hypotonia, poor suckling, and poor weight gain during infancy; early childhood-onset hyperphagia, obesity, mild mental retardation, hypogonadism, growth hormone insufficiency, and behavioural disturbances.[1] PWS patients with morbid obesity when planned for bariatric surgery present with unique anaesthetic challenges.[2,3]

A 14-year-old boy (weight-81.5 kg, height-141 cm, BMI 40.99 kg/m^2) with PWS and morbid obesity was planned for laparoscopic Roux-en-Y gastric bypass anastomosis [Figure 1]. He was born at 33 weeks of gestation, with low-birth weight and poor cry. He had delayed milestones and poor weight gain up to 09 years of life, after which he developed hyperphagia, rapid weight gain and severe snoring in sleep. Obstructive sleep apnoea (OSA) was diagnosed, with an apnoea hypopnea index (AHI) of 134.2. The lowest recorded oxygen saturation at room air was 80% while awake and 26% during sleep. Pulmonary function tests showed severe restrictive disease (FVC-21%, FEV1-26%, FEV1/FVC-117). Continuous positive airway pressure (CPAP) was started at 15 cm H_2O one week before surgery. Child was co-operative despite his low IQ. He slept in between conversation and was hyperactive when awake. His vitals were normal with room air SpO_2 88–93% and basal rhonchi in bilateral chest. Airway examination showed modified mallampati grade III with double chin. Baseline blood work was normal. Room air arterial blood gas showed pH-7.35, pCO_2-60.9 mmHg, pO_2-30 mmHg, HCO_3-29 mmol/l. ECG and 2D-Echo was normal. Child had subclinical hypothyroidism (TSH-9, T4-7.7, T3-1.9) and was not on any medication. Salbutamol nebulization was started a day prior to surgery to treat basal ronchi. Sedative premedication was avoided in view of the OSA. High risk informed parental consent for ICU stay and post-op mechanical ventilation was taken.

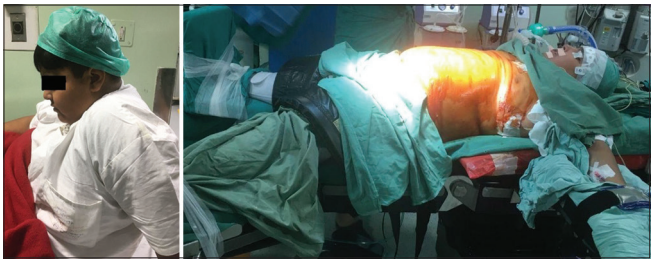

Figure 1: Patient in sitting position and after positioning

On the morning of surgery, chest auscultation showed normal vesicular breath sounds with resolution of ronchi. In the operating room, 20G intravenous (IV) line was secured. Child was placed on a ramp and ECG, pulse oximeter, non-invasive blood pressure cuff, skin temperature probe, neuro-muscular monitoring probe, bi-spectral index (BIS) monitors were attached. Pre-oxygenation with 100% O_2 was done for 03 mins. Fentanyl 100 mcg, propofol 100 mg, and succinylcholine 80 mg was administered IV. An oropharyngeal airway (size 3) was used to facilitate bag-mask ventilation. C-MAC™ guided intubation was done, using a size of 6.5 cuffed endotracheal tube (ETT) and was confirmed with auscultation and capnograph. Anaesthesia was maintained with O_2, air, desflurane (BIS 45–50). Cisatracurium was titrated to keep train of four (TOF) 0–1. Paracetamol 750 mg IV and fentanyl boluses of 20 mcg IV (03 boluses over 02 hrs) was given for analgesia. Mechanical ventilation was achieved using pressure controlled ventilation (P_{insp} 25–30 mm Hg, PEEP 7 mmHg, respiratory rate 14–18/min, FiO_2-0.5 and tidal volume 330–360 ml) with end-tidal CO_2 maintained between 40 to45. Total operative time was 02 hrs with blood loss of 100 ml. Vitals were stable throughout the procedure. Desflurane was stopped after deflation of pneumo-peritoneum. On return of spontaneous ventilation (TOF 4), neuromuscular blockade reversal was done. Even after MAC reduced to 0.2, BIS was still reading 70–75 and child did not become awake. Hence extubation was delayed for 30 mins until patient became fully awake (BIS >90) and was responsive to commands. Post extubation, child was observed for a while on the OT table and then shifted to postanaesthesia care unit (PACU) with CPAP. There were no episodes of significant or persistent desaturation (<88%) and child was sent to ward after three hours.

Perioperative concerns include an obese patient with reduced pulmonary reserve, difficult airway, obstructive sleep apnea, arrhythmias, cor-pulmonale, diabetes mellitus, altered thermo-regulation, and behavioural issues. Metabolic changes like

deficiencies of protein, vitamins, iron, calcium may also be associated. These patients have a high risk of perioperative morbidity and mortality.[4]

Nafiu *et al*.[5] demonstrated that obese children were more likely to have difficult mask ventilation, difficult intubation, and prolonged PACU stay due to high incidence of postoperative airway obstruction.

To conclude, patients with Prader–Willi syndrome pose a significant challenge to the anaesthesiologist owing to obesity-associated physiological changes primarily a difficult airway, aspiration risk and cardio-respiratory instability. Aggressive and defiant behavior often adds to the difficulties. A thorough pre-operative workup by the anaesthesiologist, endocrinologist, pulmonologist, and paediatrician for identification and optimization of co-morbidities would go a long way in planning for perioperative management.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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