Original Research Paper

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Internal Medicine

ANAESTHETIC MANAGEMENT OF A CHILD WITH PRADER WILLI SYNDROME WITH RIGHT UNDESCENDED TESTIS FOR RIGHT ORCHIDOPEXY

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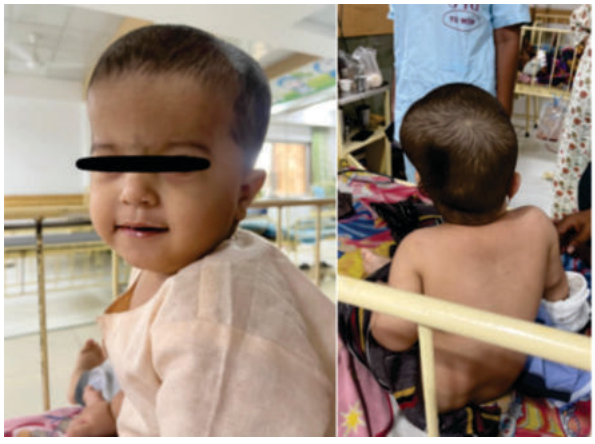
ABSTRACT Prader Willi syndrome is a rare genetic disorder characterized by hypothalmopituitary abnormalities, severe hypotonia, risk of morbid obesity and delayed psychomotor development which presents with unique anesthetic challenges. We report a case of 2 year old male child k/c/o Prader Willi syndrome posted for right orchidopexy under general anesthesia.

KEYWORDS : Prader Willi, Pediatric Age Group, Psychomotor Development, General Anesthesia.

INTRODUCTION

Prader Willi syndrome is genetically heterogenous and caused by anomalies in chromosome 15(15q11-q13). The child presents with morbid obesity, craniofacial abnormalities, sleep apnea, risk of respiratory failure, altered thermoregulation and glucose intolerance(1). PWS is a two-staged disorder with a hypotonic early infantile phase and an obese childhood phase. The infantile phase is characterized by failure to thrive, delayed milestones, seizures, fair skin and eyes. The later stage is characterized by obesity due to increased appetite along with behavioural changes, hypotonia, hypogonadal, dental caries, skeletal abnormalities, skin picking and spontaneous bruising (2).



CASE REPORT

Preoperative

A 2 year male child, weighing 12.5 kilograms k/c/o Prader willi syndrome with global developmental delay, hypotonia and cranial vault deformity. Patient had h/o NICU stay for 15 days after birth i/v/o hypoglycemia, hypotonia and hypoxic ischaemic encephalopathy, came with complaints of right undescended testis since 1.5 years.

Airway assessment showed large tongue, retrognathia, cranial deformity, retrognathia present; vitals heart rate-110bpm oxygen saturation-98% on room air. On auscultation S1S2 heard, no murmurs and bilateral air entry equal, no added sounds. Laboratory investigations within normal limits. 2D echo revealed normal study MRI brain- Cerebral atrophy and dilatation of ventricles with corpus callosum thinning.

Intraoperative

All standard ASA attached, preoxygenation with 100% oxygen for 3 minutes with infant mask size 1. Premedication with injection glycopyrrolate 0.05mg IV and induced with injection midazolam 0.25 mg IV, injection fentanyl 25mcg injection propofol 25mg IV injection atracurium 6.25mg IV and muscle relaxation status was monitored with train of four (TOF), injection dexamethasone 1.25mg, injection hydrocortisone 25mg, injection paracetamol 250mg

Patient was intubated with 4.5 sized uncuffed endotracheal tube using laryngoscope blade 2. Tube was fixed at 13 cm, bilateral air entry confirmed, fresh gas flow oxygen and nitrous at 2 litres per minute, sevoflurane started. Endotracheal tube secured and put on pressure control mode (TV-116, RR-22, Fi02-50%, PEEP-5). Surgery was uneventful. Analgesia was provided with hernia block 0.25% Bupivacaine 5ml intraoperatively. After the surgery TOF ratio was 0.85. Patient was extubated after reversal with glycopyrrolate 0.1mg and neostigmine 0.625mg.

Post extubation patient had irregular breathing and desaturation.Nasal airway inserted and patient put in left lateral position .Mask ventilation was done with 100% oxygen for 20 minutes with intermittent assessment of respiratory efforts. Saturation improved from 85% to 100% on oxygen. Nebulisation done with Budecort and Duolin respules. Injection Deriphyllin 1mg/kg intramuscularly given. After monitoring for 30 minutes and Aldrete score 9/10 patient was shifted to post op recovery followed by 24 hours observation, no desaturation observed.

DISCUSSION

Prader Willi syndrome is genetically heterogenous and caused by anomalies in chromosome 15(15q11-q13) (1).

Both general and regional anesthesia can be challenging as using general anesthesia may result in difficult airway management and landmarks for regional anesthesia may be obscured due to morbid obesity (2,3). Anesthesia concerns being morbid obesity, sleep apnea, difficult IV access, potential for difficult airway, risk of respiratory failure, myocardial involvement, aggressive behavior, seizures thermoregulation and glucose intolerance (4).

Prader Willi syndrome patients are predisposed to gastric aspiration due to hypotonia and the sequel of aspiration may be more severe due to limited pulmonary reserves. So, efforts should be made to reduce gastric secretions, increase intestinal motility, neutralize, stomach, contents, secure the airway and extubate cautiously (3).

Using general anesthesia with a nondepolarizing muscle relaxant can be a difficult due to potential for long lasting neuromuscular blockade if hypotonia is present. Thus, neuromuscular block monitoring was done in our patient using TOF (5).

Airway management may be complicated by the frequent association of poor dentition, hypotonia and palatal abnormalities which was seen in our patient post operatively, bronchospasm for which 20 minutes mask ventilation was done leading to improvement in saturation followed by nebulization in the recovery (6).

CONCLUSION

Though rare in occurrence, the patients with PWS can pose significant anaesthetic challenges mandating a thorough preoperative evaluation and management plan. The spectrum of complications such as difficult venous access, obesity, airway changes, respiratory complication, metabolic changes and OSA should be kept in mind. Our patient had PWS with undescended testis, where in right orchidopexy was performed successfully under general anaesthesia.

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