Letters to Editor

Anesthesia for percutaneous nephrolithotomy in a case of Kartagener’s syndrome

Dear Editor,

Kartagener’s syndrome is an autosomal recessive disorder characterized by clinical triad of situs inversus including dextrocardia, bronchiectasis, and sinusitis.\textsuperscript{[1,2]} We report the anesthetic management of a patient with Kartagener’s syndrome posted for percutaneous nephrolithotomy (PCNL) under general anesthesia.

A 25-year-old male was admitted to the hospital with pain in the left flank and burning micturition. He also had history of repeated respiratory infections, chronic sinus infections, and bronchospasm. Chest X-ray showed dextrocardia, cystic bronchiectasis predominantly in left base and stomach bubble on right side. Ultrasound abdomen showed situs inversus. Both X-ray Kidney Ureter Bladder film and ultrasound abdomen revealed a staghorn calculus in left kidney with moderate hydronephrosis. Electrocardiogram (ECG) demonstrated sinus rhythm with lead I displaying inversion of P wave, deep Q wave and inversion of T wave. V1 to V6 leads demonstrated progressively decreasing amplitude of R wave. The ECG was normal when all the leads were reversed and the 2D echocardiography was also normal. Pulmonary function tests revealed FEV\textsubscript{1} (Forced Expiratory Volume in one second) of 65% of predicted, FVC (Forced Vital Capacity) of 91% of predicted and FEV\textsubscript{1}/FVC ratio of 73%. Spirometry showed improvement after bronchodilator therapy. Pre-operative arterial blood gas analysis showed pH 7.33, PaO\textsubscript{2} 87 mm Hg, PaCO\textsubscript{2} 48 mm Hg and HCO\textsubscript{3} 22 mmol/L. Based on the presence of classical triad, Kartagener’s syndrome was diagnosed during workup for the surgery. During pre-anesthetic visit, patient had cough with whitish viscous expectoration for which he received vigorous chest physiotherapy and postural drainage. He was also given nebulization with salbutamol and fluticasone twice a day. Parenteral antibiotics and incentive spirometry were started pre-operatively.

The patient was shifted to operation theatre for left PCNL under general anesthesia. Intraoperative monitoring included ECG with all leads reversed, pulse oximetry, noninvasive blood pressure, temperature, capnography and neuromuscular monitoring. Midazolam 2 mg and inj. fentanyl 2 µg/kg IV were given as premedication. He was pre-oxygenated and anesthesia induced with propofol 2.5 mg/kg IV. Two minutes before intubation, intravenous lignocaine was given in order
Letters to Editor

we used propofol for intubation and to suppress airway reflexes. After muscle relaxation with succinylcholine, the trachea was intubated. Anesthesia was maintained with oxygen, air and isoflurane with vecuronium as neuromuscular blocker. The end-tidal CO2 was maintained between 30 mm Hg and 35 mm Hg with the tidal volume of 10-12 ml/kg, Respiratory Rate of 10/min with I: E ratio of 1:2. Airway pressures were lower than 25 mm Hg and oxygen saturation was 100%. Humidification was maintained by heat and moisture exchanger. After intubation, patient was placed in prone position for surgery. Good hydration was maintained with crystalloids (Normal saline and ringer lactate solution) 2.2 L. Total duration of surgery was 2 h and 20 min and consisted of two kidney punctures including one supracostal puncture. At the end of surgery, two nephrostomy tubes were placed and analgesia was given with infiltration of nephrostomy tract with bupivacaine 0.5% 20 ml solution. Neuromuscular blockade was reversed with neostigmine 0.05 mg/kg and atropine 0.6 mg IV. Immediate post-extubation, oxygen saturation dropped to 93% which improved with throat suctioning with the help of laryngoscope. Paracetamol and ondansetron were given once in 8 h post-operatively.

Incentive spirometry and chest physiotherapy was started in the postanesthesia care unit (PACU). On 2nd post-operative day, patient developed an episode of bronchospasm with no new changes in repeat X-ray chest, which was treated with intravenous theophylline. Patient improved, shifted to the ward and was discharged on 5th post-operative day after removal of nephrostomy tubes.

Kartagener’s syndrome is a variant of immotile cilia syndrome which is an autosomal recessive disorder of microtubules of ciliated epithelial cells.1 Because of the impairment of ciliary motility, patient with Kartagener syndrome can present with respiratory difficulty in the peri-operative period. Primary goals in the management would be assessment of cardiopulmonary function and prevention of pulmonary complications.1 Our patient presented with repeated chest infections. Pre-operatively, thorough chest physiotherapy, postural drainage, inhaled bronchodilators, inhaled steroids and antibiotics helped. Patients with dextrocardia reveal a sinus rhythm when ECG leads are reversed as in our patient. When dextrocardia occurs with situs inversus, heart is mostly normal, without any evidence of congenital heart disease as in our case.1

In patients with history of bronchospasm, the incidence of wheezing is higher when thiopental is given for induction than in those given propofol.3 The mechanism of the relative bronchodilating effect of propofol is not known. In our patient, we used propofol for intubation and to suppress airway reflexes prior to intubation intravenous lidocaine was given 2 min before tracheal intubation. Humidification was done with heat and moisture exchanger and good hydration was maintained to decrease the viscid secretions.

It is important to prevent lung complications by prevention of post-operative nausea and vomiting and good perioperative analgesia, in addition to proper suctioning in the perioperative period.2 Whenever possible local or regional anesthesia is to be preferred over general anesthesia.4 In our patient, because of the staghorn calculus in the renal pelvis, general anesthesia was planned. Furthermore, there is nasal cavity narrowing by chronic sinusitis in Kartagener’s syndrome, hence oropharyngeal airway is preferred over the nasal airway as was used in our case post-operatively.

Ventilatory parameters with large tidal volumes (10-15 ml/kg) combined with slow inspiratory flow rates as used in our case minimize the likelihood of turbulent airflow and help maintain optimal ventilation-to-perfusion matching.5 Slow respiratory rates (6-10 breaths/min) provide sufficient time for complete exhalation to occur.

In summary, the anesthesiologist should be aware of the peri-operative complications and implications of cardiopulmonary inversion in patients of Kartagener’s syndrome.

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Dear Editor,

We recently had a patient shifted to our tertiary care intensive care unit (ICU) via air travel. The patient was spontaneously breathing with oxygen by facemask. He had chest tube in situ and had no signs of respiratory distress prior to airlift, however, during the air travel in a medically equipped helicopter his condition worsened (desaturation up to 85% with tachypnea) and he required endotracheal intubation with ventilatory assistance, however after landing he improved within minutes and his requirement for mechanical ventilation vanished. The above finding prompted us to examine possible causes of his unique condition of spontaneous worsening and improvement.

The literature on management of patients with pneumothorax and chest drains is scarce. Patients are often not optimally managed prior to air travel and clinical condition worsens in air, where management resources are limited. We discuss the physics and physiological management involved for better outcomes of such patients.

Medical helicopters fly around 500-1000 feet above the ground while commercial/medical airplanes fly at 24-40 thousand feet above the ground level. The plane cabins are often pressurized and atmosphere barometric changes may be minimal, depending upon the efficacy of pressurizing. Presuming the temperature of flight is maintained as at the ground level, thus by “Boyle’s Law” the volume of gas is inversely related to its pressure. So as the flight ascends the atmospheric pressure falls and the volume of gas trapped in closed body cavity would expand. Normal sea level pressure is 760 mmHg and on ascent to 8000 feet it falls to 560 mmHg (a 25% reduction), this would mean that air in cavities expands by 25%. Another aspect important in these patients is the decrease in PaO$_2$ with ascent. At sea level the partial pressure of oxygen is around 150 mmHg, on ascent it falls to around 110 mmHg (this is equivalent to use of FiO$_2$ of only 15% at sea level).

In our patient, it was later realized that although chest tube was optimally functioning, he still had loculated collection that expanded (compressing lung) and in addition the lower apparent FiO$_2$ lead to dyspnea.

As per guidelines by American College of Chest Physicians, patients with spontaneous pneumothorax should not air travel for at least a week after complete resolution on imaging. Patients with recurrent pneumothorax or predisposition should not air travel unless a chest drain or surgical intervention is done.

In patients with a chest drain, the functioning of drain must be confirmed and no clamping should be done at the time of travel. Prior to travel, imaging must be done to rule out any extra-pulmonary air not in communication with the drain. Wherever possible patients must be transported in closed pressurized chamber planes, as the artificial air ‘pumped in’ maintains both external pressure and FiO$_2$.

In our patient, the size of loculated air, on landing again decreased and FiO$_2$ increased, thus patient immediately recovered. It must be kept in mind that these problems are preventable if appropriate steps are taken.

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