Anesthetic Implications of Prone Position Percutaneous Nephrolithotomy in a Deformed DWARF
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INTRODUCTION
The physiognomy and the frequently associated physiologic abnormalities of the respiratory, circulatory and neurologic systems of dwarfs dictate they must be considered differently.¹ Genitourinary abnormalities have been reported to occur in 43% of patients with congenital scoliosis.² Patients with kyphoscoliosis are at an increased risk of urolithiasis due to abnormal kidney ascent, renal tract deviations, poorly draining urinary systems and higher incidence of urinary tract infections.³ An understanding of pathophysiology, precise anesthesia planning and modern anesthetics in these patients could increase margin of safety and improve outcome.

CASE REPORT
A 47 year male, weighing 27 kg, and height of 97.5cm presenting with intermittent bilateral flank pain since 1 to 2 years. He had severe long bone deformity (Figure-1). This condition ran in the family, as his father and children were affected too. He was also a known asthmatic on medications. He was diagnosed to have renal calculi thus posted for percutaneous nephrolithotomy (PCNL). General examination revealed a thoracolumbar kyphoscoliosis and pectus carinatum (Figure-2). He was edentulous, short necked, Mallampatti classification 3 with restricted range of terminal neck extension. He had a respiratory rate of 14/min. without use of any accessory muscle of respiration with breath holding time of 37 seconds. His room air saturation was 98%. On auscultation air entry was bilaterally equal with no adventitious sounds. Heart sounds were normal with no murmur. Routine laboratory investigations were normal. X-Ray Chest showed thoracolumbar scoliosis with unequal lung fields. Echocardiography showed mild pulmonary hypertension with good left ventricular systolic function. Pulmonary function test was unavailable to us as the machine could not be calibrated to his height and weight. Hence we only did a room air ABG which showed a pH of 7.459 with PCO₂ 31.8 mmHg, PO₂ 137 mmHg and SO₂ 99%. USG abdomen revealed mild hydronephrosis of Rt. Kidney with 1.8 cm calulus in pelvis and 5 mm calculus in lower calyx of the Lt. Kidney. Radiograph of thoraco lumbar spine showed significant osteoporosis of lumbar vertebrae with compression and fish mouth appearance which could be consistent with osteodystrophy. Preoperatively patient was optimized with adequate hydration, nebulization and chest physiotherapy. After obtaining written valid high risk consent; patient was wheeled into the operation theater. Monitoring devices instituted were pulseoximeter, cardioscope, manual non invasive blood pressure, temperature probe and end tidal CO₂ monitoring. All the drugs were administered as per Kg body weight. Patient was premedicated with Inj. Glycopyrrolate 0.1 mg IV, Inj. Midazolam 0.5 mg IV, Inj. Fentanyl 50 + 20 micro gram IV. Patient was induced with Inj. Rocuronium 30 mg was given. ETT no. 7.5 was inserted with minimal neck manipulations. Fiberoptic bronchoscope was used to adjust and confirm tube length. The tube was fixed at 17.5 cm at the lip and bilateral air entry confirmed. For PCNL, patient was log rolled to prone position over transversely placed bolsters with abdomen free. All the pressure points were well padded to protect from fracture.

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The interpretation of restrictive lung disease with reduced spirometry, blood gas, hematocrit and serum electrolyte determination as well as diffusion capacity measurements can help in quantifying the extent of underlying pulmonary involvement. The interpretation of spirometric measurement of lung volumes is complicated in dwarfs because there are no reference standards for these measurements in short trunks dwarfs. Pulmonary hypertension leading to Cor pulmonale is probably the most common cardiovascular disturbance that develops in dwarfs. Thus the anesthesia management in these patients must be planned such that the various anesthetic agents and various other stimuli that aggravate pulmonary arterial vasoconstriction can be avoided and yet adequate cardiac output and coronary perfusion pressure is maintained. Automated BP cuffs may be hazardous in dwarfs as over inflation may result in fractures. Bleeding diathesis have been known to occur from qualitative platelet abnormality as patients due to large number of them in a population. Any practicing anesthesiologist is likely to encounter dwarfs as the patient was lost to follow-up.

**DISCUSSION**

Any practicing anesthesiologist is likely to encounter dwarfs as patients due to large number of them in a population. Our patient was distinguished by the presence of bony deformity due to brittle bones in childhood which led to a disproportionate growth of his body. He had complains of bilateral renal calculi and PCNL was planned in prone position. PCNL as a monotherapy has advantages in removing larger stones and achieving excellent results with minimal recurrence and morbidity even in difficult cases such as those involving renal anatomic variation. The patient’s position is crucial for PCNL, because the area of access is limited, especially in patients with kyphoscoliosis. Prone positioning provides a larger area for percutaneous renal access and a wider space for instrument manipulation with fewer chances for visceral injuries. Hence our surgeons had chosen prone position for this surgery. General anesthesia was considered for our patient as regional anesthesia could lead to unpredictable spread of drugs. The high level of regional block required for this surgery could be detrimental to the already compromised pulmonary functions.

Our patient had normal facial features with short neck and restricted terminal neck extension. Short neck and cervical spine abnormalities could contribute to difficult laryngoscopies in dwarfs. The sternal prominence due to pectus carinatum can interfere with midline positioning of the laryngoscope. Occasional abnormalities of skull base may limit neck extension which rarely interferes with laryngoscopy and intubation in patients. The clinical impression of an anatomically abnormal upper airway or of tracheal narrowing should be confirmed preoperatively by one or more of the following studies i.e. lateral neck radiography, xerography, tomography, CT scanning or MRI technique. In our case mask ventilation and intubation was not difficult. Short handle laryngoscope may be useful. Guidelines for the selection of the appropriately sized ETT for dwarfs are unclear. It was also difficult to determine the tube depth by the common Owen and Cheney technique due to his short stature. Hence FOB was used to adjust the tube depth such that the cuff was beyond the cords and prevent endobronchial intubation. Unintentional endobronchial intubations are the most prevalent cause of oxyhemoglobin desaturation during an otherwise uneventful general anesthesia. Restrictive lung disease with reduced vital capacity and functional residual capacity characterizes thoracic dystrophy and kyphoscoliosis. These reduced lung volumes predispose towards airway closure accompanied by subsequent ventilation /perfusion(V/Q) mismatching, increased alveolar-arterial gradient and recurrent scattered atelectasis. Spirometry, blood gas, hematocrit and serum electrolyte determination as well as diffusion capacity measurements can help in quantifying the extent of underlying pulmonary involvement. The interpretation of spirometric measurement of lung volumes is complicated in dwarfs because there are no reference standards for these measurements in short trunks dwarfs. Pulmonary hypertension leading to Cor pulmonale is probably the most common cardiovascular disturbance that develops in dwarfs. Thus the anesthesia management in these patients must be planned such that the various anesthetic agents and various other stimuli that aggravate pulmonary arterial vasoconstriction can be avoided and yet adequate cardiac output and coronary perfusion pressure is maintained. Automated BP cuffs may be hazardous in dwarfs as over inflation may result in fractures. Bleeding diathesis have been known to occur from qualitative platelet abnormality in these patients.

Careful positioning of patient is required during anesthesia and surgery. Prone positioning reduces cardiac index due to reduced venous return, direct effects on arterial filling and reduced left ventricular compliance secondary to increased thoracic pressure. Mean arterial pressure (MAP) is maintained by increased systemic vascular resistance (SVR), and pulmonary vascular resistance (PVR) also is increased in the majority of patients. The choice of anaesthetic may influence the degree to which these changes occur.
There is an increase in functional residual capacity (FRC) in prone position due to reduction of cephalad pressure on the diaphragm and reopening of atelectatic segments which was advantageous in our patient. Airway resistance is not altered by change in position. The increase in FRC and alteration in the distribution of both ventilation and perfusion throughout the lung leads improved ventilation/perfusion matching and thus improved oxygenation in the patient. Tracheal compression may occur in prone position due to thoracic scoliosis. It is seen that longitudinally placed bolsters are better to avoid hypotension. In our case bolsters needed to be placed horizontally due to pectus carinatum and hemodynamics were well preserved. The airway is more protected from regurgitation in prone position. Decrease in MAP due to increased intrathoracic pressures in our patient could offset his balance, as there would be a subsequent increase in pulmonary pressures too. Recent work suggests that anaesthetic technique could affect hemodynamic variables in the prone position. A greater fall in mean arterial pressure (MAP) is seen in total i.v anaesthesia than with inhalational agents. Hemodynamic stability and early recovery is most important part of anaesthesia maintenance in these patients. Desflurane causes a decrease in blood pressure depending on concentration. It causes a transient increase in heart rate depending on the dose, which is related to the stimulation of the sympathetic system. Desflurane confers profound protection against renal ischemia-reperfusion injury compared with pentobarbital or ketamine anaesthesia by attenuating inflammation. This may have significant clinical implication regarding choice of volatile anaesthetic in already damaged kidney. Residual effects of volatile anaesthetic may impair ventilator drive of dwarfs in the recovery room. In this regard desflurane is advantageous because of their more rapid washout and their minimal effects on hypoxic sensitivity at sub anaesthetic concentration. Desflurane is a weak trigger for malignant hyperthermia. Effective analgesia will reduce the incidence of postoperative pulmonary complications. Weight adjusted appropriate drug dosages need to be given in these patients. In our case, the rotation and curvature of spine caused modifications of anatomic relationships between ribs and spine, kidneys and the surrounding organs and a limited percutaneous exposure of the renal cavities making puncture difficult and time consuming. This also increased the risk of hydropneumothorax which was ruled out intraoperatively by visualizing bilaterally clear CP angles under fluoroscopy. Also other reported complications are colonic injury, renal vascular and parenchymal injury and septicemia.

CONCLUSION

To conclude, an understanding of the multiple abnormalities that affect dwarfs will ensure the safe delivery of anesthesia in these patients. Monitoring and anaesthetic techniques for the case should be dictated by the type of dwarf, the anatomic and physiologic aberrations of different organ systems and the nature of the surgical procedure. Prone position has its advantages and disadvantages in these patients. Safe use of desflurane is possible as they offer multiple advantages in these group patients.

REFERENCES


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