Case Report

Case Report of Congenital Kyphoscoliosis with Myotonic Dystrophy Type 1: Perioperative and Anesthetic Considerations

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CASE REPORT

A 13-year-old girl weighing 25 kg with congenital kyphoscoliosis with myotonic dystrophy type 1 (DM1), delayed motor milestones, dysmorphic features, and learning disability was posted for surgery [Figure 1].

Radiographs showed thoracic kyphoscoliosis with coronal Cobb angle 64° and kyphosis 78° with T6 hemivertebrae [Figure 2]. Diagnosis of DM1 due to expansion of triplet repeats in DMPL gene was confirmed on genetic testing. EMG was not diagnostically conclusive.

Her chest radiograph, 2D echo, ECG, ultrasound abdomen, complete blood count (Hb 13.3 mg%), coagulation profile, and renal and liver function tests were done besides arterial blood gas (ABG) analysis (pH 7.4, PaO₂ 90 mm Hg, PaCO₂ 36.8, HCO₃ 24). Pulmonary function tests (PFT) showed a restrictive pattern with FEV1 29%, FVC 27%, and FEV1:FVC 10.5.

She was cooperative and was suggested spirometry exercises. Considering myopathy, restrictive lung function, and plan of intraoperative motor evoked potentials (MEP) monitoring, we planned to avoid muscle relaxants. ICU bed and ventilator was kept standby. Informed valid high-risk consent was obtained.

Patient was brought on OT table, and peripheral intravenous (iv) line was secured. Injection midazolam 0.5 mg iv, glycopyrrolate 0.1 mg iv, and fentanyl 25 µcg iv were given slowly. Although preoxygenation was given, monitors were attached for ECG, heart rate (HR), NIBP, temperature, and SpO₂, which were monitored during surgery.

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Induction was done with propofol 2–3 mg/kg with O₂+sevoflurane 2%. Endotracheal intubation was performed with no.6 portex cuffed endotracheal tube (after 4% xylocaine spray on cords) and confirmation with EtCO₂ trace and bilateral chest auscultation. Tube was secured with bite block. No depolarizing agent was used for intubation. Left radial artery cannulation for invasive blood pressure (IBP) monitoring and Foley catheterization was done. Electrodes for somatosensory (SSEP) and MEP neuromonitoring were applied by neurotechnicians. Adequate padding of eyes and care of pressure points were taken. Baseline SSEP and MEP signals were obtained before turning prone and during surgery.

Anesthesia was maintained on O₂+nitrous oxide 50:50 with sevoflurane 2%–3%. Injection fentanyl 25 µg, iv propofol infusion, and injection dexmedetomidine 0.3mcg/kg infusion were started after turning prone. The aim was to keep mean arterial pressure (MAP) 65–70 mm Hg and minimum alveolar concentration (MAC) 0.4 to avoid interference with MEP.

Tranexamic acid 250 mg bolus and infusion was given over an hour. Fluid maintenance was with crystalloid fluids 8–10 mL/kg. Blood loss of approximately 400 mL was replaced with one packed cell volume.

Two surgeons were operating simultaneously which helped in reducing anesthesia time and blood loss. T2–T10 posterior spinal osteotomies and instrumented fusion with pedicle screws, rods, and iliac autograft were performed. Dexmedetomidine and propofol infusion were shut off approximately 40 and 30 min prior to the end of surgery, respectively. Local anesthetic solution (0.25% bupivacaine) was infiltrated at site of incision and iv paracetamol and dexamethasone were given. After turning her supine, all vitals were assessed. Temperature was maintained with Bair Hugger and iv fluid warmer. Blood sugar level and ABG were sent. All anesthetic agents were closed except O₂. Duration of surgery was 3.5 h and anesthesia 5 h.

When patient was awake, maintaining SpO₂, EtCO₂, and generating tidal volume of approximately 150–200 mL, she was extubated and kept in recovery room for observation with O₂ supplementation.

Postoperative radiographs and investigations were normal. Adequacy of respiration was monitored with respiratory rate and SpO₂. The patient was asked about pain, requirement for analgesia, and leading questions for anything she remembered about surgery and discharged 5 days postoperatively after an uneventful recovery. At 1 year postoperative visit, she was well balanced, with correction maintained. [Figures 3, 4] Thoracic kyphosis improved by 48% (38°) and scoliosis by 78% (49°).
**Discussion**

Congenital myopathies and muscular dystrophies are rare conditions.\(^1,2\) Muscular dystrophies comprise of DM1, DM2, Duchenne muscular dystrophy, and fascioscapulohumeral dystrophy.\(^3\) DM1 (Steinert disease) has an incidence of 2.1–14/100,000, and is an autosomal dominant disorder with congenital, childhood, or adult onset.\(^2\) Multisystem involvement of cardiac, pulmonary, central nervous, endocrine, ocular, and musculoskeletal systems with varying severity may occur.\(^2-4\) Patients with myopathies and muscular dystrophies are susceptible to cardiopulmonary complications, rhabdomyolysis, and malignant hyperthermia (MH).\(^1-3,5\) These patients need a detailed history about muscle tissue disorders and anesthetic complications in other relatives over several generations to avoid missing underlying disease while doing preanesthetic fitness; moreover, identification often poses a challenge due to subclinical or late onset of symptoms, as mutations occur de novo and positive family history may be absent.\(^1-3\) Congenital scoliosis is the result of congenital vertebral abnormalities and may be associated with cardiac, renal, and tracheoesophageal anomalies.\(^6,7\) Thoracic Cobb angle ≥60° is associated with a decrease in PFT.\(^6-8\) DM1 and thoracic scoliosis are associated with restrictive PFT with difficulties in airway management.\(^1-4,6-8\)

Myopathic patients should be first to be anesthetized as workstations must be free of volatile anesthetic remnants as they are susceptible to MH.\(^1\) Tight temperature control is essential for prevention of hypothermia as it prolongs anesthesia and interferes with neuromonitoring.\(^1,2,9\) Preoperative sedatives and opioids should be used with caution as they may cause respiratory insufficiency.\(^1-3\) Airway should be managed with a modified rapid sequence induction and depolarizing agents like Succinylcholine, must be avoided in cases of DM1 as it can cause muscle contractions and MH.\(^1-3\) Inhalation agents, sevoflurane, and nitrous oxide-O₂ have been used uneventfully.\(^2\) Propofol infusion, fentanyl and nondepolarizing agent atracurium, have been used in a series of 13 DM1 patients without adverse effects.\(^3\) Non-depolarizing agents should be avoided for scoliosis as they interfere with MEP. Dexmedetomidine has been used as an adjunct in TIVA with minimal effect on MEP and SSEP.\(^9\) Deliberate hypotension and hypothermia may result in neuromonitoring changes.\(^9\)
Intraoperative and postoperative blood loss is increased in myopathies, neuromuscular scoliosis, and with spinal osteotomies.[1,10] Blood loss can be minimized by careful positioning, tranexamic acid, two surgeon approach. Conservation strategies like isovolemic hemodilution, cell salvaging should also be considered.[10]

Due to the high prevalence of co-morbidities, myopathic patients require intense postoperative monitoring in ICU. 26.5% patients with certain myopathies affecting cardiac and respiratory system experience postoperative complications.[6] Screening for myoglobinuria should be done to detect early signs of MH and anesthesia related rhabdomyolysis.[1,2,5] Mechanical ventilation should be continued if not fully awake or if severe gas exchange abnormalities.[2,8] Optimization of pulmonary status is achieved by aggressive chest physiotherapy. Paracetemol and NSAIDs are safe for analgesia, while opioid infusions can be used with caution with monitoring.[2]

**Conclusion**

Congenital spinal deformity with underlying DM1 is rare. Curve progression and deteriorating pulmonary function are surgical indications. Patients are susceptible to cardiopulmonary complications, rhabdomyolysis, and malignant hyperthermia. Anesthesia management, measures for reducing blood loss, and operative time with adequate postoperative monitoring are emphasized.

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**Conflicts of interest**

There are no conflicts of interest.

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