A novel approach to the anaesthetic management of a case of osteogenesis imperfecta

Sir,

Osteogenesis imperfecta (OI) is a rare connective tissue disorder which manifests in approximately 1 in 20,000 births. Manifestations include short stature, short neck, blue or grey sclera, defective dentition, brittle bones, coagulopathy, hypermobile joints, restrictive pulmonary disease and craniovertebral junction anomalies. Anaesthesiologists usually encounter these patients for orthopaedic surgeries either to correct deformities or to fix fractures. OI always imposes challenges for the attending anaesthesiologists due to susceptibility for new fracture during shifting or positioning, difficult airway, difficult regional anaesthesia, platelet dysfunction, vulnerability to malignant hyperthermia and other associated systemic comorbidities.[1-4]

Here, we report a case of OI tarda with difficult airway who underwent general anaesthesia uneventfully for internal fixation (nailing) of fracture shaft femur where endotracheal intubation (ETI) was executed with a GlideScope™ (Verathon, Bothell, WA, USA) video-laryngoscope and a novel 180° upside down technique was used. In this case, parents did not consent for regional anaesthesia.

A 17-year-old female known case of OI tarda weighing 22 kg and 98 cm in height was scheduled for nailing of fracture shaft femur. General physical examination revealed features of OI including blue sclera, impaired dentition, short neck, pectus carinatum, thoracic scoliosis and bent long bones. Airway examination revealed mouth opening of 2.5 cm, modified Mallampati Class 3, short neck and defective dentition. The patient was accepted for surgery as American Society of Anesthesiologists (ASA) Grade III patient. ASA standard monitoring which includes electrocardiography, capnography, pulse oximetry, temperature and urine output was done. Baseline blood pressure was 116/70 mmHg, heart rate...
was 92/min and oxygen saturation on room air was 96%-97%.

After preoxygenation, general anaesthesia was induced with fentanyl 50 µg and propofol 50 mg intravenously. Mask ventilation was assured and atracurium 12 mg was administered intravenously to facilitate airway instrumentation. In view of difficult airway, GlideScope™ video-laryngoscope was chosen for ETI. It was difficult to insert the blade of GlideScope™ into patient's mouth with manual in-line stabilisation of the cervical spine. Gentle jaw thrust enabled video-laryngoscopy. Cormack-Lehane Grade IIa glottis view was obtained, but ETI was not successful. Then, 180° upside-down manoeuvre was used for ease of endotracheal tube (ET) insertion. The ET was inserted into the mouth with convexity anteriorly and tip facing towards the hard palate with manufacturer's stylet (GlideRite™) in situ. ET was then rotated 180° anticlockwise and gently advanced in such a way that the curvature followed the contour of floor of mouth till it approached the glottic aperture [Figure 1]. GlideRite™ was withdrawn 2.5–5 cm outside and ET was further slid into trachea.

Anaesthesia was maintained with oxygen, air, propofol (100 µg/kg/h) and fentanyl (1 µg/kg/h) infusions with atracurium boluses. Pressure controlled mode of ventilation was used. At the end of surgery, reversal of neuromuscular blockade was done with neostigmine and glycopyrrolate. Postoperatively, the patient was monitored in post-anaesthesia care unit for 1 h. The patient was discharged from the hospital on the sixth post-operative day.

OI's hallmark is the mutation in the gene (COL1A1 and COL2A2) that leads to either defective collagen formation or reduction in collagen formation, causing bones to become brittle and prone for fractures. Overextension of the cervical spine during direct laryngoscopy can cause odontoaxial dislocation and vertebral artery compression.[7] ETI with fibre-optic bronchoscope is the preferred method in such cases.[8] In our case, difficult airway anticipation was made due to short neck, mouth opening of 2.5 cm, large tongue, Mallampati Grade III and pigeon chest deformity. Fibre-optic bronchoscope was not available, and the patient was not an ideal candidate for intubating laryngeal mask airway (weight <30 kg). Video-assisted laryngoscopy is recommended as an initial approach to intubation in the management of difficult airway.[9] Cervical spine was manually stabilised in line by a second assistant to prevent odontoaxial dislocation: An attempt was made to insert the GlideScope™ blade in patient’s mouth but failed due to the large tongue and pigeon chest deformity. Gentle jaw thrust which pulled the tongue forward enabled the insertion of GlideScope blade in patient’s mouth and successful ETI was executed using the 180° upside-down technique.[7] We would like to emphasise that manual in-line cervical spine stabilisation and gentle jaw thrust and the 180° upside-down technique of ET insertion by GlideScope™ video-laryngoscope are alternative techniques of airway management that can be used in OI cases.

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Conflicts of interest
There are no conflicts of interest.

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REFERENCES
1. Oakley I, Reece LP. Anesthetic implications for the patient with osteogenesis imperfecta. AANA J 2010;78:47-53.
2. Widmann RF, Bitan FD, Laplaza FJ, Burke SW, DiMaio MF, Schneider R. Spinal deformity, pulmonary compromise, and quality of life in osteogenesis imperfecta. Spine (Phila Pa 1976) 1997;22:1380-6.
3. Edge G, Okafor B, Fennelly ME, Ransford AO. An unusual manifestation of bleeding diathesis in a patient with osteogenesis imperfecta. Eur J Anaesthesiol 1997:14:215-9.
4. Rampton AJ, Kelly DA, Shanahan EC, Ingram GS. Occurrence of malignant hyperpyrexia in a patient with osteogenesis imperfecta. Br J Anaesth 1984:56:1443-6.
5. Rodrigo C. Anesthesia for maxillary and mandibular osteotomies in osteogenesis imperfecta. Anesth Prog 1995;42:17-20.
6. Apfelbaum JL, Hagberg CA, Caplan RA, Blitt CD, Connis RT, Nickinovich DG, et al. Practice guidelines for management of the difficult airway: An updated report by the American Society of Anesthesiologists Task Force on Management of the Difficult Airway. Anesthesiology 2013:118:251-70.
7. Singh M, Kumari K, Kapoor D, Singh J. “180° upside down maneuver” for ease of endotracheal tube insertion with the GlideScope in patients with limited mouth opening. J Clin Anesth 2013:25:243.