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CLINICAL VIGNETTE

Perioperative Management of a Patient with Osteogenesis Imperfecta Undergoing Orthopedic Surgery

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Background

Osteogenesis imperfecta (OI) is an inherited connective tissue disorder that occurs in one in every 15,000-20,000 births.¹ It is characterized by increased susceptibility to fractures in the setting of minimal or no trauma. Patients may also have scoliosis, osteopenia, chest wall deformities, poor dentition, and increased incidence of valvular heart defects.^{1,2}

The genetic cause of OI is increasingly thought to be polygenetic, with mutations affecting the transcription, translation, or post-translational modifications of Type 1 collagen, a principle component of bone and cartilage. The majority of cases, however, are caused by an autosomal dominant mutation affecting the gene encoding the COL1A1 and COL1A2 pro-peptides of type 1 collagen.³ To minimize the morbidity and mortality associated with this disease, patients are often treated by an interdisciplinary team of providers with treatment options including surgery, pharmacotherapy, and physical therapy.

Case Report

A 21-year-old female with a history of osteogenesis imperfecta was scheduled for an open reduction and internal fixation (ORIF) of a right bicondylar tibial plateau fracture that was sustained after a ground level fall. Patient was diagnosed with OI as a young child, and was not on any OI medications prior to presenting for surgery. Her past medical history was also notable for well-controlled asthma and impaired glucose tolerance. Prior surgeries included repair of a right patella fracture under general anesthesia with no reported problems with anesthesia.

On physical exam, the patient had a normal heart and lung exam, and a body mass index of 29 kg/m². Her airway exam revealed a Mallampati score of 3, decreased thyromental distance of 2 finger breadths, full range of motion of her neck, and normal mouth opening. Of note, she had multiple missing and broken teeth with several loose lower teeth. A preoperative ECG, echocardiogram, and pulmonary function testing showed no evidence of cardiopulmonary dysfunction. Additionally, cervical spine radiographs were significant for "straightening of cervical lordosis" which was stable with flexion and extension of the neck. Laboratory studies including complete blood count, basic metabolic panel, and coagulation studies were all within normal limits.

On the day of surgery, the patient was taken to the operating room and placed on standard American Society of Anesthesiology monitors, including EKG, non-invasive blood pressure cuff, pulse oximetry, and capnography. The patient was positioned carefully in the supine position, keeping the head in a neutral position, and ensuring that all pressure points were padded. General anesthesia was induced with fentanyl, lidocaine, and propofol, and the patient was paralyzed with rocuronium. Intubation was performed with a video-assisted laryngoscope, keeping the patient's neck in a stable position throughout. A size 7.0 endotracheal tube was placed uneventfully. Esophageal temperature probe was used to monitor core body temperatures. The case proceeded uneventfully, and she was extubated and transferred to the post-anesthesia care unit in stable condition at the end of the surgery.

Discussion

Osteogenesis imperfecta is a diverse condition with many clinical manifestations that both increase the likelihood that a patient with OI may require surgery and complicate their anesthetic management. The specific considerations depend both on the severity and type of manifestations in these patients, and therefore, a thorough preoperative evaluation is essential.

One of the most important intraoperative considerations for patients with OI is their increased risk of fracture with minimal trauma. As a result, extra precaution should be taken when moving and positioning these patients. If possible, positioning should be done prior to induction of anesthesia so the patients can provide feedback on any concerning pressure points or discomfort. Additionally, the compressive force exerted by traditional blood pressure cuffs may be enough to cause a fracture in severe cases.⁴ In this case, non-invasive continuous blood pressure monitoring may be preferable over the standard cuff. However, a recent retrospective study showed no association of NIBP monitoring with bony injuries and recommended against the routine use of invasive arterial lines for hemodynamic monitoring in OI patients.⁵ Furthermore, new technology such as the EV1000 provides non-invasive blood pressure monitoring without the use of a traditional blood pressure cuff.

Patients with OI also have specific considerations regarding their airway. Given their predisposition for fragile dentition

and possibility of vertebral fracture with manipulation of the cervical spine, particular care must be taken during intubation. This challenge is compounded by the fact that patients may also have accelerated osteoarthritic changes of their cervical spine, limiting their neck extension and ability to achieve an adequate view during intubation. In this case, we opted to use a video-assisted laryngoscope for the intubation to ensure that her cervical spine neutrality was maintained and that her dentition remained intact. Some anesthesiologists have recommended the use of an intubating laryngeal mask in patients with OI.⁶ If neck extension is severely limited and there is high concern for dental or vertebral damage, fiberoptic intubation may be considered as well.

Spinal anesthesia may also be more difficult in patients with OI. The increase in osteoarthritic changes and the increased incidence of scoliosis in OI patients make traditional anatomic landmarks less reliable and the level of the block more difficult to predict.

Additionally, it has been reported that patients with OI may have a predisposition to hyperthermia during anesthesia administration and in the immediate postoperative period. The specific mechanism of this phenomenon has not been fully elucidated but is thought to be distinct from malignant hyperthermia.^{7,8} It has been recommended to minimize the use of warming blankets and some reports recommended using total intravenous anesthesia (TIVA) rather than inhalational anesthetics.

Conclusion

Osteogenesis imperfecta is a rare and heterogeneous clinical disorder with clinical manifestations both requiring surgical correction and potential complicated anesthetic management during these procedures. We present a young adult with OI who underwent general anesthesia for a complication of her condition. The diversity of clinical presentations of OI can make it difficult to predict the specific challenges that may arise during anesthesia, but understanding the potential complications is essential for safe and effective anesthetic administration for these patients.

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