

ANESTHETIC MANAGEMENT OF A PATIENT WITH OSTEOGENESIS IMPERFECTA: A CASE REPORT



Mihaela Visoiu, M.D., Banu Lokhandwala, M.D., Jean Charchaflied, M.D.

Department of Anesthesiology, SUNY Downstate Medical Center, Brooklyn, New York

BACKGROUND

Osteogenesis imperfecta (OI) is an inherited connective tissue disorder often called "brittle bone disease". It is a rare disorder with an estimated incidence of about 1/20,000. It is associated with multiple skeletal and systemic manifestations that present many anesthetic challenges.

CASE PRESENTATION

We describe the anesthetic management of 65 yo female, with a history of OI, (type severe tarda), undergoing left radical total parotidectomy with facial nerve resection and reconstruction for squamous cell carcinoma. Her clinical manifestations included severe kyphoscoliosis and growth stunting, multiple limbs contractures and deformities (**Figure 1**), and restrictive pulmonary disease. In addition, she had hypertension and left ventricular failure, with ejection fraction of 20%. The patient was very knowledgeable and concerned about her medical condition, and expressed that by advising the medical team about the importance of special padding and positioning to avoid fracture or other skeletal injury.

The medical team responded by reassuring the patient and incorporating her wishes into the care plan. She was allowed to move herself to the operating table by using her sliding board, a technique that she was familiar with. Then she positioned herself on the operating table with her arms and legs fully padded. No strap was applied. The blood pressure (BP) site on the upper extremity was well padded and the BP cuff was measured only as needed before and during induction. Before induction, midazolam, and fentanyl were given to alleviate anxiety and discomfort. Anesthesia was induced using propofol, fentanyl, followed by rocuronium, to facilitate endotracheal intubation, which was performed with extra care to avoid any pressure to the skeletal or soft tissue structures in the mouth and pharynx. After induction, an intra-arterial catheter was inserted in the right radial artery, and BP cuff was disconnected. Anesthesia was maintained using infusions of propofol and remifentanyl, in addition to isoflurane. Emergence from anesthesia was smooth and the patient was extubated in the operating room, and transported to the recovery room with monitors

Figure 1:
Osteogenesis Imperfecta (severe tarda)



Commonly seen malformations may include: severe osseous fragility (brittle bones), severe deformity of long bones, kyphoscoliosis with chest deformity (pectus excavatum) growth retardation with short stature, ligamentous laxity, dentinogenesis imperfecta, wormian bones (small irregular bones along the cranial sutures)

Figure 2:
Examples of Blue Sclera



Figure 3:
Dentinogenesis Imperfecta



Marked discoloration and severe attrition of the primary dentition are seen in this child with dentinogenesis imperfecta type II. *Pediatr Clin North Am* 2000, 47:975

and oxygen. The patient was observed in the recovery room for four hours, and was eventually discharged home four days after surgery.

DISCUSSION

OI present challenging anesthetic issues such as perioperative fractures and dislocation, difficult intubation, respiratory complications due to kyphoscoliosis and pectus excavatum, excessive bleeding due to platelet dysfunction, severe intra-operative metabolic acidosis, latex allergy, eye injury from external pressure, hyperhydrosis, aortic and mitral valve dysfunction, and intra-operative hyperthermia. However, there is no conclusive evidence of association of OI with Malignant Hyperthermia.² Additional manifestations in OI include bluish sclera (**Figure 2**) in infant, defective dentition (**Figure 3**), premature deafness, hydrocephalus and brain stem compression due to cranial development defects.

CONCLUSIONS

There is no ideal anesthetic recipe for patients with OI and no specifically contraindicated agents. Successful anesthetic management requires comprehensive pre-operative assessment and preparation, followed by meticulous intra-operative and postoperative care. Patients with OI often require multiple procedures and having the same anesthesiologist for subsequent procedures may enhance both the anesthesiologist's and the patient's sense of comfort.

REFERENCES

1. Marini, JC: Osteogenesis imperfecta: comprehensive management. *Adv Pediatr* 1988, 35:391.
2. Hall RMO, Henning RD, Brown TCH and Cole WG: Anaesthesia for children with osteogenesis imperfecta: A review covering 30 years and 266 anaesthetics. *Paediatric Anaesthesia* 1992, 2:115-121.