

Introduction

Osteogenesis Imperfecta (OI) is a brittle bone condition inherited genetically at birth. Gene variation can occur from spontaneous mutation, paternally, maternally, or both depending on the type of OI¹. There are several different severities arising mainly from type 1 collagen defects that impact bone metabolism². OI patient's bones are at an increased risk of fracture, and further complications include scoliosis, respiratory infections, heart problems (valvular malfunction), hearing loss, blue sclera, soft discolored teeth, CNS abnormalities, and vision loss³. Diagnosis can be confirmed through lab tests and x-ray. The main anesthetic goal is to prevent deformities and fractures during intubation and positioning.

Type	Inheritance	Severity	Fractures	Bone Deformity	Stature
I	AD	Mild	Few to 100	Uncommon	Normal or slightly short for family
II	AD	Perinatal lethal	Multiple fracture of ribs, minimal calvarial mineralization, platyspondyly, marked compression of long bones	Severe	Severely short stature
III	AD Rare recessive	Severe	Thin ribs, platyspondyly, thin gracile bones with many fractures, "popcorn" epiphyses common	Moderate to severe	Very short
IV	AD	Moderate to mild	Multiple	Mild to moderate	Variably short stature
V	AD	Moderate	Multiple with hypertrophic callus	Moderate	Variable
VI	Uncertain	Moderate	Multiple	Rhizomelic shortening	Mild short stature
VII	AR	Moderate	Multiple	Yes	Mild short stature

AD: Autosomal dominant, AR: Autosomal Recessive

Figure 1: Types of OI⁴

Significance/Objectives of Case

It can be quite rare to provide anesthetic care to a patient with OI in the adult population because only a predicted 25,000-50,000 people in the U.S. have this disease⁴.

1. Increase awareness, knowledge, and expectations regarding patients with OI.
2. Review anesthetic considerations for the OI patient population.
3. Provide intraoperative care insight for OI management.

Patient Description

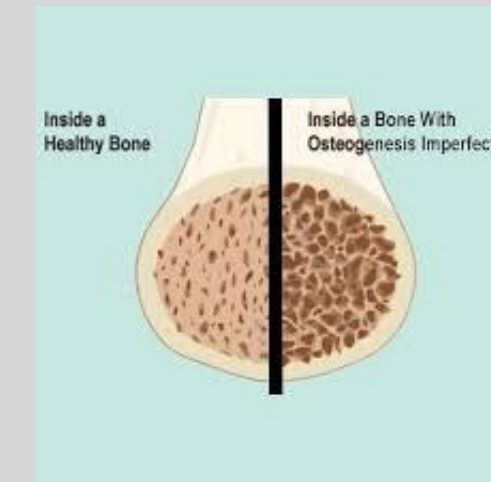
A 36yo F with nephrolithiasis presented for right percutaneous nephrolithotomy and ureteral stent placement. A BMI of 27.71kg/m² and height of approximately 99cm (3'3") were recorded. Relevant medical history included OI, scoliosis, and PONV. The patient was unaware of which type of OI she was diagnosed with; however, based on clinical assessment and dependence on a motorized wheelchair for transportation, she most likely had type IV. The patient's only known allergy was latex. She did not take any notable medications. The patient reported daily recreational inhaled marijuana use, minimal alcohol consumption, and no tobacco use. Past surgical history included numerous orthopedic procedures for fractures, dental work, spinal fusion, and percutaneous nephrostomy. All her surgeries had been uneventful, with the last one being 4 years prior. Her anesthetic records indicated an easy mask, and she was successfully intubated with direct laryngoscopy using a MAC 3 blade and 7.0mm ETT.

Intervention/Anesthetic Method

- IV access was difficult due to patient's limited extremity movement and comfort. Ultimately, a 22g IV was placed in the patient's L forearm.
- Patient was moved to OR bed via undersheet ensuring whole body movement and no strain on her extremities.
- NIBP monitoring was doable with this patient on her forearm. Some patients can't even have a BP cuff on since the strain of the cuff squeeze can cause bone fractures. Arterial lines should be considered.
- Gentle BMV with no neck manipulation was performed to avert strain on cervical spine.
- Videoscope intubation (glidescope) with LoPro S3 blade, rigid stylet, and 6.0mm ETT, secured at 18cm after confirming bilateral breath sounds.
- Patient was carefully positioned in L lateral decubitus, with extra padding on pressure points.
- A 20g IV was placed in R forearm after positioning
- TIVA was performed to reduce PONV incidence.

Images

Mouth Opening: 4-5 cm
Mallampati: II
TM distance: 4-5 cm
C spine ROM: Limited Extension and Limited Flexion
TMJ Jaw Protrusion: Incisors can meet
Neck: Thick / Obese



Left, above, and below: Examples of bone deformities in OI⁵



Discussion

Although the operation took longer than expected because of the patient's abnormal anatomy, anesthetic management was uneventful and successfully performed. BMV was easily attained. Intubation was more complicated as the patient's airway was slightly deviated to the right, which was corrected with gentle cricoid pressure. Vigilance was applied around the patient's teeth. BLBS were checked to ensure avoidance of mainstem intubation. TV was reduced to 280mL with less PEEP. The patient was carefully placed in L lateral decubitus with additional padding. A second IV was placed. PONV was not present postoperatively, and no damage was done.

Conclusion

Depending on the severity of OI, changes to the anesthetic plan must be made. It is important to ensure that all those providing perioperative care to these patients are aware of the need for careful patient movement/positioning and ensuring adequate padding on all pressure points. Consider videoscope intubation to prevent cervical spine manipulation, and arterial line- especially if procedure is expected to take longer. Succinylcholine is also avoided, if possible, to avoid muscle fasciculations that could cause fractures. Encountering OI in clinical practice is rare. However, being aware of these patients and their special considerations will ensure safe and effective care.

References

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