Anesthesia for Maxillary and Mandibular Osteotomies in Osteogenesis Imperfecta

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A 21-yr-old female suffering from osteogenesis imperfecta was anesthetized for correction of maxillary and mandibular deformities that had restricted her chewing. Preoperative assessment revealed a difficult intubation, restrictive lung disease secondary to bony deformities, and multiple repairs of fractures. Management of anesthesia for this operation—which is very rarely carried out in this disorder—is described along with a review of the problems of anesthesia associated with osteogenesis imperfecta.

Osteogenesis imperfecta is a rare disorder of the connective tissues. Elective jaw operations in osteogenesis imperfecta are rarely carried out due to the increased bone fragility in these patients. This case report describes the anesthetic management for such an operation in a patient who had osteogenesis imperfecta.

CASE REPORT

A 23-yr-old female with type I osteogenesis imperfecta and associated dentinogenesis imperfecta was referred to maxillofacial surgery with a complaint of difficulty in chewing. Following a diagnosis of maxillary hypoplasia anteroposteriorly and vertically and mandibular hyperplasia anteroposteriorly, the surgeons decided to perform a maxillary Le Fort I advancement and down grafting, together with a mandibular body step osteotomy, removing a premolar bilaterally and using the premolar edentulous space for the set back of the mandible. The patient was then referred for anesthetic assessment.

The patient's hearing was normal. Thus, there was no problem in communication. Her past medical history revealed multiple fractures of all limbs, mainly the lower. Most had been treated conservatively, but some had required surgical intervention. She had osteotomies and pins inserted to both tibia and had an open reduction of a supracondyler fracture and fixation with K wires, all under general anesthesia without complications. However, some delay in bone healing had been reported. The last general anesthetic was 9 yr ago. There was no clear family history of this condition.

She weighed 26.5 kg. On examination she was found to have the characteristic frontal and parietal bone protrusion together with an overhanging occiput. Her sclerae were blue. She had a severely prognathic mandible that restricted opening of the mouth because it impinged on her prominent sternum ("pigeon chest"). She also had kyphoscoliosis. Her neck was short, and neck movements were restricted anteroposteriorly by the severe kyphosis of the spine and the pigeon-shaped anterior chest. Her upper anterior teeth were missing. The legs were deformed, with limited extension. Thus, she could not walk more than a few steps and was confined to a wheel chair. She could not sleep flat due to the marked curvature of the spine. Her skin was pale, thin, and translucent.

Investigations found the electrocardiogram, blood picture, and coagulation tests to be normal. Platelet function tests were not carried out as there had been no problems with hemostasis previously. Echocardiography did not reveal any valvular disease. Blood chemistries, apart from mild increases in calcium, phosphates, and alkaline phosphatases, were normal. Respiratory function tests revealed a severe restrictive defect, with a forced vital capacity of only 1.2 L and a 1-sec, forced expiratory volume of 1.08 L (Table 1). Oral radiography indicated marked bone loss in the mandible and features of dentinogenesis imperfecta, namely opalescent dentine, curved roots, and partially obliterated pulp canals.

The patient was premedicated with 7.5 mg of diazepam orally and was made to rest on a molding mattress, which enabled her to lie supine. Her preoperative body temperature was normal. In the induction room, monitoring of her blood pressure, electrocardiogram, pulse,

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Table 1. Preoperative Respiratory Function Tests by Spirometry

Test	*Units*	*Predicted*	*Actual*	*% Predicted*
FVC	L	1.94	1.20[a]	62
FEV_1	L	1.90	1.08[a]	57
FEV_1/FVC	%	98	89	91
$FEF_{25-75\%}$	L/sec	3.18	1.51[a]	47
$FEF_{50\%}$	L/sec	3.53	1.84[a]	52
$FEF_{75\%}$	L/sec	2.72	0.78[a]	29
PEF	L/sec	6.27	3.40[a]	54
FIVC	L	1.94	1.07[a]	55

Abbreviations: FVC, forced vital capacity; FEV_1, forced expiratory volume in 1 sec; $FEF_{25-75\%}$, forced mid-expiratory flow; $FEF_{50\%}$, forced expiratory flow at 50% exhalation; $FEF_{75\%}$, forced expiratory flow at 75% exhalation; PEF, peak expiratory flow; FIVC, forced inspiratory vital capacity.

[a] Outside normal range.

and oxygen saturation was commenced. She had an inhalational induction with nitrous oxide, oxygen, and halothane. Once induced, a 16-ga cannula was inserted into a vein in her left arm, and an infusion of Hartmann's solution was started. Through this cannula she received 2.5 mg of dexamethasone and 25 μg of fentanyl. Following spraying the nasal passages with 4% cocaine, a 28-ga reinforced Rusch latex endotracheal tube was introduced nasally into her trachea with the aid of a fiberoptic bronchoscope. Through the other nasal passage a 16-ga nasogastric tube was inserted into the stomach, and the position checked. Then, the cuff of the endotracheal tube was inflated to occlude the trachea, and the throat was gently packed with a ribbon gauze soaked in saline and wrung dry by hand.

She was transferred to the operating table on the molding mattress. She was positioned in a reverse Trendelenberg position to facilitate venous drainage. With the additional monitoring of end-tidal carbon dioxide and temperature, she was paralyzed with 3 mg of vecuronium, and her lungs were ventilated using a Narkomed IV ventilator (North American Drager, Telford, PA) with nitrous oxide, oxygen, and isoflurane in a semiclosed circuit with a circle absorber. Increments of fentanyl (6.25 μg) and vecuronium (0.5 mg) were given hourly. One mega unit of crystalline penicillin was given intravenously as antibiotic cover.

Following injection of 2% lidocaine with 1:80,000 epinephrine into the mucosa of the maxilla, surgery of the maxilla was commenced. In order to reduce bleeding, the blood pressure was maintained between 80 and 100 mm Hg with the aid of 0.25-mg increments of propranolol. Once the maxillary segments were in place, the mandibular step osteotomy was carried out after the additional injection of local anesthetic. The surgeons were especially careful not to lean on the patient, and no heavy instruments were placed on the body.

All vital signs remained within normal limits throughout the procedure. Temperature increased by about 1° C during the surgery. The total blood loss was approximately 300 mL. She was given 250 mL of plasma protein in addition to the clear fluids.

About half an hour before the end of the procedure, she was given 3.125 mg of prochlorperazine intramuscularly. At termination of surgery, the action of the muscle relaxant was reversed with 1.25 mg of neostigmine and 0.6 mg of atropine. The throat pack was removed, and once her respiration was sufficient to maintain normal oxygen saturation and an end-tidal carbon dioxide tension of less than 50 mm Hg, she was extubated. She coughed during and after extubation, indicating the presence of protective reflexes. As she did not have any respiratory obstruction following extubation, she was transferred to the recovery room.

The patient was monitored in the recovery room and given 30% humidified oxygen. She recovered uneventfully and was transferred to the ward. The postoperative period was uneventful.

DISCUSSION

Although the anesthetic course was uneventful and appeared to be normal, this was due to the proper assessment, preparation, and management of the case—essentials in treating patients whom many anesthetists may come across once in their career or never at all. Osteogenesis imperfecta is a rare inherited connective tissue disorder that primarily affects bone. The basic defect appears to be due to a specific mutation within the type I collagen genes.[1]

Our patient was anesthetized by us when she was an adult. However, like our patient, many individuals with osteogenesis imperfecta present for anesthesia during childhood. Although our patient had no loss of hearing, 10% may have deafness, thus making communication difficult.[2]

Osteogenesis imperfecta leads to many anesthetic problems. For example, bony abnormalities may cause difficulties. A hanging occiput[2] and/or a short neck may limit extension of the neck, making intubation difficult. The prominent protruding mandible may limit mouth opening if it is associated with a pigeon chest and a short neck, as in our case. The mandible may be brittle,[3] also as was seen radiologically in our case, leading to the possibility of mandibular fracture during intubation. Fragile, brittle teeth, which are a feature of dentinogenesis imperfecta, can easily be damaged or dislodged during intubation. Some of the teeth may be missing, which may cause the laryngoscope blade to slip into the cleft, which in turn may damage the remaining teeth or make intubation difficult. Occasionally, the presence of a cleft pal-

ate[2] may result in similar problems during intubation. Excessive extension of the cervical spine during difficult intubation may result in cervical spine fracture due to the fragility of the bones. If one is experienced with the fiberoptic bronchoscope, it appears to be the ideal aid for intubation in these patients.

Past history of absence of anesthetic complications, especially during childhood, should not deter proper preoperative investigation. As the child grows to an adult, abnormal growth of bones, loose teeth, and absence of teeth can cause intubation problems.

Due to the brittleness of bone, succinylcholine is best avoided,[4] as fasciculations produced by it may result in bone fractures. Deep inhalational anesthesia, as used in our case, or a short-acting nondepolarizing muscle relaxant like ropivacaine, may be used to relax the patient during intubation.

For short operations, mask anesthesia may be difficult in these patients due to the abnormal facial anatomy. When using a mask, one must be careful not to fracture the brittle mandible. A laryngeal mask may help to maintain the airway. However, insertion of the laryngeal mask should be done gently to avoid dislodging or damaging the teeth and causing fracture of the mandible.

Patients with osteogenesis imperfecta may have kyphosis, scoliosis, pectus carinatum (pigeon chest) or pectus excavatum,[5] or a combination of these. Furthermore, they usually present with limb abnormalities and a history of multiple fractures. Joint laxity may result in joint dislocation. These physical liabilities often cause problems when positioning the patient. As seen in our case, the patient may be unable to lie supine. A molding mattress should be used to position the patient. If one is not available, movement of the patient should be done with extreme care, and pressure points should be padded to protect the excessively thin and fragile skin. The surgeons should be warned not to lean on the patient or place heavy instruments on the patient.

Respiratory function is often compromised due to abnormalities of the chest wall, such as the kyphoscoliosis seen in our case.[6] Significant respiratory impairment may result in cor pulmonale.[3] It is essential to assess respiratory function preoperatively and monitor it carefully both intra- and postoperatively. Patients with osteogenesis imperfecta may not be able to compensate for respiratory obstruction due to their weak chest wall. Dexamethasone should be given preoperatively in elective correction of facial deformities to reduce the postoperative swelling, which may compromise the airway.

Cardiac abnormalities, such as mitral or aortic incompetence, atrial or ventricular septal defect, or patent ductus arteriosus, may be a problem with these patients.[7] Clinical examination and echocardiography can be helpful in detecting these abnormalities.

Bleeding may also be a problem with these patients, because of the type of surgery performed and/or platelet dysfunction.[8] Tests may be carried out preoperatively to assess platelet function. In our case, platelet function tests were not performed because the patient's coagulation times were normal and she had undergone operations before without experiencing excessive bleeding. Good positioning to promote venous drainage and mild hypotensive anesthesia, as carried out in our case, may facilitate surgery in these patients.

There are many reports[4,5,9,10] of increases in temperature in these patients, both with and without anesthesia. The hyperthermia may be due to an associated increase in the metabolic rate of these patients.[11] It may be accompanied by an excess of thyroid hormone secretion. Preferably preoperatively, the thyroid hormone concentrations should be determined. It is advisable to postpone elective operations if the patient has an increased temperature preoperatively.[5] Patients with this disease who have received hyoscine[12] or atropine[11] have higher temperatures preoperatively and during anesthesia. Thus, in these patients it is advisable to use anticholinergic drugs only when they are specifically indicated. Our patient had a normal body temperature preoperatively, and the temperature rose by 1° C during the operation, probably from heat retained under the drapes. Pulse oximetry and end-tidal carbon dioxide monitoring may herald a warning in case of hypermetabolism during surgery. The increase in temperature is said to be a nonmalignant hyperthermia.[13,14] One should be ready to cool the body if the temperature rises. The use of a water blanket, cold intravenous solutions, and a decrease in the operating room temperature should suffice. If the body temperature rises, an increased oxygen concentration to compensate for the increased oxygen consumption and adjustment of ventilation to maintain normocarbia should be carried out. Increased fluids should be infused to compensate for the perspiration losses.

An antiemetic should be given prophylactically to prevent vomiting postoperatively, as the act of vomiting may dislodge the repairs and/or fracture the brittle facial bones.

Finally, these patients, though deformed, are normally intelligent and verbal. They are aware of their potential mortality. Thus, they should be provided appropriate emotional support in addition to supportive physical handling.[4]

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