

Anaesthetic Management of a Case of Osteogenesis Imperfecta with Bladder Outlet Obstruction: A Case Report

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ABSTRACT

Osteogenesis Imperfecta (OI) is a fibro-osseous disorder of the collagen tissue which leads to defects in the skeletal growth and short stature. OI poses various anaesthetic challenges, which include difficult intubation, problems with positioning and a tendency to develop malignant hyperthermia, coagulopathy and cardiovascular abnormalities. We describe the anaesthetic management of an 18 year old boy, a known case of OI with kyphoscoliosis, who presented with acute retention of urine due to urethral stone and was successfully managed for

emergency suprapubic cystolithotomy with caudal anaesthesia. He had characteristic features like short stature, short neck, blue sclera, a receding mandible, pectus carinatum (pigeon chest), hyper mobile joints, kyphoscoliosis and a history of recurrent hospitalization for long bone fractures. Since our patient presented for an emergency surgery, we had only little time to evaluate and plan out the management, which posed significant challenges. A successful outcome was ensured by the carefully obtained history, examination and gentle care, with the application of basic principles in managing this patient.

Key Words: Osteogenesis imperfecta, Emergency surgery, Caudal epidural, Urethral calculus

INTRODUCTION

Osteogenesis Imperfecta (OI), also known as brittle bone disease, is a fibro-osseous disorder of the collagen tissue which leads to defects in the skeletal growth and short stature. The defect in the skeletal growth is a result of the lack of normal ossification of the endochondrial bone, resulting in an increased fragility of the bones [1]. The anaesthetic implications of OI include difficult intubation, problems with positioning and a tendency to develop malignant hyperthermia, coagulopathy and cardiovascular abnormalities [2]. We describe the anaesthetic management of an 18 year old boy, a known case of OI with kyphoscoliosis, who presented with acute retention of urine due

to a urethral calculus and was successfully managed by emergency suprapubic cystolithotomy with caudal anaesthesia.

CASE REPORT

An 18-yr-old boy, a known case of OI, presented to the emergency room with acute retention of urine. Foley's catheterization was unsuccessful. The x-ray of the pelvis showed a urethral calculus and he was taken up for urethroscopy. On pre-anaesthetic evaluation, the patient was found to weigh 20 kg, he had a height of 106 cm and he was bed ridden. He had characteristic features [Table/Fig-1] like short stature, short neck, blue sclera, a receding mandible,



[Table/Fig-1]: Photograph of the patient showing skeletal abnormalities

pectus carinatum (pigeon chest), hyper mobile joints, kyphoscoliosis and a history of recurrent hospitalization for long bone fractures, for which he was treated conservatively. His younger sibling was also a known case of OI. On assessment of the airway, he was found to have adequate mouth opening, normal dentition and acceptable neck movements and he had the airway assessment classification, Mallampatti grade-II. His spine examination revealed kyphoscoliosis. His vital parameters and other systems were within normal limits.

Investigations revealed haemoglobin = 9.5 g%, bleeding time = 3 min 30 sec, clotting time = 4 min, platelet count = 22 5000/mm³, blood sugar = 96 mg/dl, blood urea = 35 mg/dl, serum creatinine = 1.2 mg/dl, serum sodium = 138 meq/l, serum potassium = 4.2 meq/l and serum calcium = 8.4 mg/dl. His chest x-ray, ECG and echocardiogram were normal. The x-ray of his spine showed dorsolumbar kyphoscoliosis. Regional anaesthesia was planned. A written informed consent was taken. In the operating room, after securing intravenous access with a 20G cannula, routine monitors including ECG, non-invasive blood pressure and pulse oximetry were applied. A difficult airway cart was kept ready. Under aseptic precautions, caudal epidural anaesthesia was administered with 15 ml of 0.25% bupivacaine in the lateral position and a T₁₀ level of block was achieved. With utmost care, he was put in a lithotomy position and his pressure points were padded. Intravenous Midazolam 0.5 mg and Fentanyl 25 ug were administered as sedation. Suprapubic cystolithotomy was performed as the urethral calculus was pushed into the bladder. His vital parameters remained stable intra-operatively and the procedure lasted for about 40 min. The patient had an uneventful recovery and was discharged on the 8th post-operative day.

DISCUSSION

Osteogenesis Imperfecta is a rare inherited disease of the connective tissue, which occurs in two forms, Osteogenesis Imperfecta Congenita and Osteogenesis Imperfecta Tarda. With the congenita form, death usually occurs in-utero [1]. The tarda form typically manifests during childhood or early adolescence, but the patients have a normal life span. The prevalence of OI ranges from 1:60,000 to 1:20,000, depending upon the type. Its inheritance generally follows an autosomal dominant pattern, although sporadic cases are common [2].

These patients usually present with blue sclera, recurrent fracture of the bones, hyper mobile limbs, and other associated skeletal deformities like kyphoscoliosis, shortening, pigeon chest, etc. The bleeding disorder is common due to platelet dysfunction. These children can have associated cardiovascular and neurological abnormalities [3]. They might present with obstructive uropathy following renal and ureteric stones [2].

As our patient presented for an emergency surgical procedure, we had only little time to investigate and prepare for the procedure. However, a careful history, assessment and examination guided us to rule out any associated problems and thereby to provide safe anaesthesia. There have been several successful case reports of the conductance of surgery under general as well as regional anaesthesia in patients with OI. Karabiyik et al have used total intravenous anaesthesia (TIVA) along with intubating laryngeal mask airway to manage a patient who underwent nephrolithotomy and ureterolithotomy, while Malde et al. have successfully used balanced general anaesthesia in case of OI for abdominal hysterectomy [1,3,4].

Due to abnormal skeletal growth, the risk of odontaxial dislocation and hyper-mobile joints, a difficult airway must always be anticipated in these patients. In our patient, we avoided general anaesthesia due to an anticipated difficult airway, lower abdominal surgery and susceptibility to malignant hyperthermia. Bergstrom in 1977 and Rampton et al in 1984 have reported several cases of malignant hyperthermia in the OI patients [5,6]. The use of succinylcholine should be avoided because of its potential to cause malignant hyperthermia and because of the occurrence of fractures due to fasciculations. The fragility of bones in these patients is well known. As excellently highlighted by Vogel et al, the positioning difficulties and the management of these problems call for a detailed consideration of those abnormalities [7]. Malde et al have reported the fracture of the shaft of the femur in a patient of OI, which occurred during the transfer of the patient to the recovery room [4]. Intra-operatively, utmost care was taken while positioning the patient for the lithotomy.

Increased intra-operative bleeding may occur despite the normal bleeding times and the coagulation values. Increased capillary fragility, decreased levels of factor VIII and deficient platelet aggregation have been implicated as the causes for this. In our patient, before administering regional anaesthesia, a quick pre-operative work up was done, with special attention to the coagulation profile, which was within normal limits. Caudal epidural was chosen as the preferred anaesthesia technique over spinal anaesthesia, due to an anticipated difficulty in performing a subarachnoid block due to kyphoscoliosis and due to the unpredictability of the level of the block. To summarize, a successful outcome was ensured by carefully obtaining the history, by examination and by gentle care with the application of basic principles in managing this patient.

In conclusion, with this experience, we would like to emphasize the need for a detailed pre-operative evaluation and for the preparation for anaesthesia. In a patient of osteogenesis imperfecta, for emergency surgeries, special attention is required to rule out associated cardiovascular abnormalities, bleeding disorders and difficult airways. An extra gentle care is essential during the positioning and the transfer of these patients.

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FINANCIAL SUPPORT AND CONFLICTS OF INTEREST

There are no external funding, financial interests and conflicts of interest involved in this study.

Date of Submission: **Mar 19, 2011**

Date of per review: **Apr 20, 2011**

Date of acceptance: **May 2, 2011**

Online first: **May 10, 2011**

Date of Publishing: **Jun 13, 2011**