A successfully managed case of Brittle Bone Disease and associated abnormalities.

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Citation

M Varshney, P Varshney, L Gupta, P Bhaduria. *A successfully managed case of Brittle Bone Disease and associated abnormalities.*. The Internet Journal of Anesthesiology. 2008 Volume 22 Number 1.

Abstract

Osteogenesis imperfecta, a connective tissue disorder, is not only characterized by brittle bones but there are other systemic involvements which should be taken into account before anaesthetizing the patient. We share our experience of anaesthetizing a case of Osteogenesis imperfecta managed successfully by taking the necessary precautions.

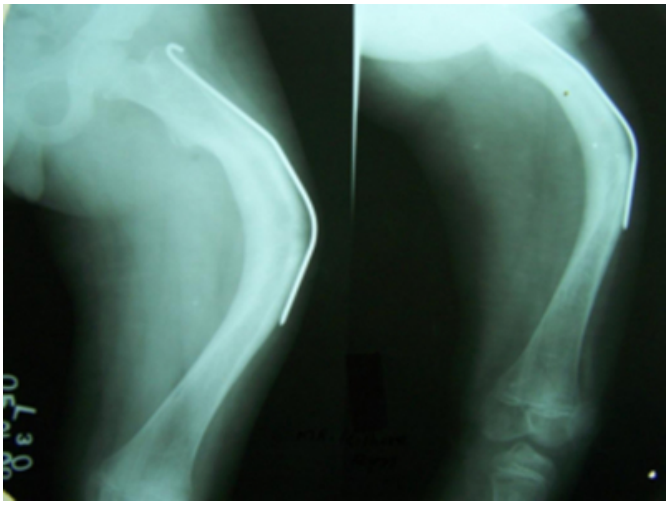
Sir,

Osteogenesis imperfecta also known as brittle bone disease is an inherited disorder of connective tissue caused by mutations in one of the two genes encoding collagen type 1. The primary bone lesion is the lack of normal ossification of the endochondral bone resulting in easy fragility of bones but there are other systemic involvements which make management of anesthesia in these patients challenging. We share an experience of anesthetizing a patient with OI posted for corrective osteotomy & internal fixation of left femoral deformity.

A 12 year old male child weighing 30 kg, known case of Osteogenesis imperfecta, was posted for corrective osteotomy & internal fixation of shepherd crow deformity of left femur (Fig 1).

Figure 1

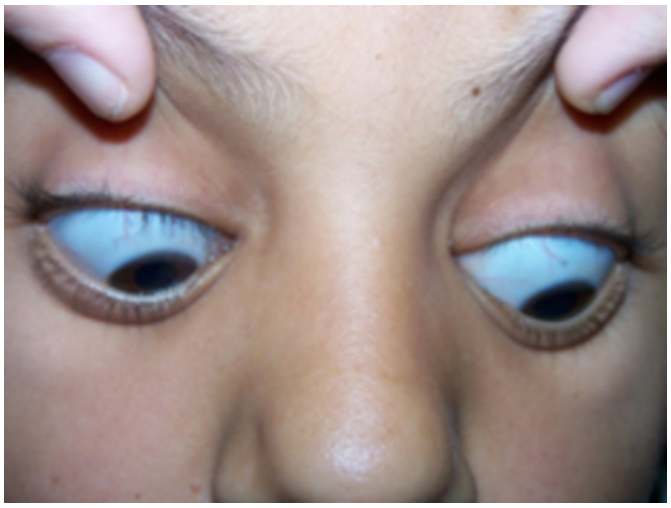
Fig 1: Shepherd crow deformity of the femur



The child gave history of multiple fractures involving both lower limbs since early childhood. Intramedullary nailing had already been done for fracture left femur around 2 year back under general anesthesia. Past history revealed delayed milestones. No other siblings were found to be similarly involved. Examination of the child revealed vitals with in normal limits, short stature (Height 119 cm), large head with frontal bossing, crowding of teeth and blue sclerae besides deformity of left thigh. Airway examination was normal. No other systems were clinically involved.

Figure 2

Fig 2: Blue Sclera



Laboratory investigations revealed normal haemogram, KFT, serum Na+ & K+. Serum Ca2+ was 7.6 mg% which improved to 9.8 mg% after calcium supplementation. Serum free T_3 (3.58 pg/ml) was slightly out of range (1.45-3.48 pg/ml) while free T_4 was with in normal limits. There was increased translucency of posterior ends of 2nd -7th ribs on chest X-ray. ECHO, done to rule out any valvular involvement, was found to be normal.

The surgery was scheduled to be performed in right lateral position. Decision was taken in favour of general anesthesia which was induced using Inj Fent 60µg & Inj Thiopentone 125 mg while the patient breathed 3% sevoflurane in O_2 & N_2O. Inj Vecuronium bromide 3.0 mg was administered for neuromuscular block. Size 2.5 Proseal was inserted with his head in neutral position. ECG, SPO_2, $ETCO_2$, Temperature and noninvasive manual blood pressure were monitored. All the limbs were adequately padded and care was taken while making lateral position to prevent any iatrogenic fractures. Anesthesia was maintained using 1% sevoflurane in O_2 (33%) & N_2O (66%) and muscular relaxation continued using Inj vecuronium bromide. Intraoperative vitals were stable. After completion of surgery the child was made supine with utmost care and anesthesia reversed using 100% O_2, Inj neostigmine 1.5 mg & Inj glycopyrrolate 0.3 mg. The patient was shifted to PACU. Rest of the postoperative course was uneventful.

OI is characterized by defect in the endochondral ossification and follows autosomal dominant pattern of inheritance though some recessive patterns have been seen. Four major clinical criteria are defined to diagnose OI (1) Osteoporosis with abnormal fragility of bones (2) Blue Sclerae (3) Dentinigenesis imperfecta (4) Premature otosclerosis. Presence of two of these abnormalities confirms the diagnosis. Various other abnormalities associated with OI include 10-30% incidence of bleeding diathesis and increased risk of surgical bleeding. The cause is platelet cell dysfunction due to abnormal collagen1. Also there is an increased incidence of aortic and mitral valve dysfunction due to dilation of valve ring. Both of these abnormalities were ruled preoperatively in our patient by appropriate investigations. Due to increased fragility of bones there is an associated enhanced risk of fractures of mandible and cervical spine during laryngoscopy & neck extension. Also due to abnormal skeletal growth, difficult airway must be expected in these patients2. Presence of dentinogenesis imperfecta makes teeth more prone to injury during laryngoscopy. Literature refers to successful use of laryngeal mask airway and intubating laryngeal mask airway in these patients3. We used Proseal LMA in our patient which was inserted in neutral head position using the introducer tool. Also in this manner drain tube was used to pass temperature probe in lower esophagus for most accurate temperature monitoring. Position of the patient is another area of concern especially lateral position as in our case. To prevent any fractures all the limbs were adequately padded and axillary role of cotton was inserted to minimize pressure in axilla and on ribs. Utmost care was practiced to prevent torsion of spine lest it should cause vertebral fractures and/or dislocation. The use of automated blood pressure cuff may be hazardous as overinflation may cause fractures. Similarly succinylcholine induced fasciculation can fracture bones so must be avoided. Most of patients with OI have mild hyperthermia with hyperhidrosis either because of abnormal central temperature regulating mechanism or due to abnormal cellular metabolism. Cooling blankets and cold iv fluids should be available to manage hyperthermia4. Also extra care should be practiced while transferring the patient to and from operating suit and to recovery room. Though we administered general anesthesia to this patient the technique of choice for such patients is conduction block as it avoids the necessity of airway manipulation, risk of hyperthermia and facilitates detection of thyroid storm. In our case the child was not cooperative enough to permit neuraxial blockade.

To conclude we would like to emphasize the need for thorough preoperative evaluation to identify other associated abnormalities in a patient of OI and to prevent and appropriately manage any untoward complication extra vigilance and preparation should be practiced.

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