

# Intracranial epidural hematoma in osteogenesis imperfecta

A large [intracranial epidural hematoma](#) can develop with [osteogenesis imperfecta](#) (OI) after a trivial [head trauma](#). Factors influencing the development are peculiar to this rare disorder of bone and connective tissue development and include bony weakness, increased vascular fragility, and possible coagulopathy <sup>1)</sup>.

Although some patients can be treated nonoperatively, other require craniotomy for clot evacuation, Surgery may be complicated in some children because of the underlying bone fragility and bleeding diathesis commonly observed in patients with [osteogenesis imperfecta](#). <sup>2)</sup>.

## Case reports

### 2013

The anesthesia management of [osteogenesis imperfecta](#) (OI) patients should be exercised with caution given certain risks of respiratory disorders. These risks are due to thorax deformity, bone fractures during moving or changing position, mandibular and [cervical fractures](#) related with intubation, difficult intubation and [malignant hyperthermia](#). The anesthetic technique using Total Intravenous Anesthesia (TIVA) and laryngeal mask airway is suitable for pediatric patient care with OI. However, these techniques have not yet been reported as useful in neurosurgery case reports. In this study, we present the use of TIVA and ProSeal Laryngeal Mask in a child with OI and epidural hemorrhage.

Erdoğan et al. came to the conclusion that LMA and TIVA can safely be used in the anesthetic management of OI patients with severe anesthetic problems <sup>3)</sup>.

### 2008

Sasaki-Adams et al. sought to determine the neurosurgical implications of OI in a cohort of patients treated at a quaternary care center for pediatrics. They reviewed the case histories of 10 children with OI treated by the neurosurgical service at the Hospital for Sick Children in Toronto between January 1988 and March 2007. The cases of 4 of these children are highlighted in the article. The most common neurosurgical conditions encountered in this cohort included macrocephaly in 5 patients, subdural hematoma in 3 patients, **epidural hematoma in 2 patients**, and hydrocephalus in 3 patients. Basilar invagination and spinal fractures were observed in 20% of the cohort. Although some patients could be treated nonoperatively, several required craniotomy for clot evacuation, decompression, and spinal fixation for fracture or basilar invagination, and cerebrospinal fluid shunt insertion. Neurosurgical conditions affecting patients with OI include macrocephaly, the development of an acute intracranial hematoma after often minimal trauma, the development of chronic subdural fluid collections that may require drainage, hydrocephalus (both communicating and noncommunicating), basilar invagination, and subaxial spinal fractures. Surgery may be complicated in some children because of the underlying bone fragility and bleeding diathesis commonly observed in patients with OI <sup>4)</sup>.

## 2007

A large epidural hematoma that developed in a child with OI after a trivial fall highlights the importance of close monitoring in these patients. After an injury that occurred several months prior to the head trauma the authors describe, this child had been placed in foster care because it was believed that his skeletal injuries were caused by nonaccidental injury. Subsequent genetic analysis confirmed that the child was heterozygous for the missense mutation c767G>T,pG256V at exon 16 of COL1A2, consistent with OI, and the foster care order was overturned <sup>5)</sup>.

## 1985

A case of an extensive extradural haematoma as a complication in a patient with osteogenesis imperfecta is reported. The significance of careful appraisal and post-traumatic, neurosurgical observation of patients with osteogenesis imperfecta and a history of recent, albeit seemingly trivial <sup>6)</sup>.

## 1983

Acute bilateral extradural hematomas occurred after apparently trivial trauma in a patient suffering from osteogenesis imperfecta congenita. Factors influencing the development of this dramatic complication are peculiar to this rare disorder of bone and connective tissue development and include bony weakness, increased vascular fragility, and possible coagulopathies <sup>7)</sup>.

<sup>1)</sup> <sup>5)</sup>

Parmar CD, Sinha AK, Hayhurst C, May PL, O'Brien DF. Epidural hematoma formation following trivial head trauma in a child with osteogenesis imperfecta. Case report. J Neurosurg. 2007 Jan;106(1 Suppl):57-60. PubMed PMID: 17233315.

<sup>2)</sup> <sup>4)</sup>

Sasaki-Adams D, Kulkarni A, Rutka J, Dirks P, Taylor M, Drake JM. Neurosurgical implications of osteogenesis imperfecta in children. Report of 4 cases. J Neurosurg Pediatr. 2008 Mar;1(3):229-36. doi: 10.3171/PED/2008/1/3/229. PubMed PMID: 18352768.

<sup>3)</sup>

Erdoğan MA, Sanli M, Ersoy MO. Anesthesia management in a child with osteogenesis imperfecta and epidural hemorrhage. Braz J Anesthesiol. 2013 Jul-Aug;63(4):366-8. doi: 10.1016/j.bjane.2012.07.008. Epub 2013 Aug 13. PubMed PMID: 24565246.

<sup>6)</sup>

Diaz LA, Lippe K. Acute extradural haematoma following trivial trauma in a case of osteogenesis imperfecta. Neurochirurgia (Stuttg). 1985 Jul;28(4):180-1. PubMed PMID: 4033853.

<sup>7)</sup>

Pozzati E, Poppi M, Gaist G. Acute bilateral extradural hematomas in a case of osteogenesis imperfecta congenita. Neurosurgery. 1983 Jul;13(1):66-8. PubMed PMID: 6877569.

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