

PBLD #11 MODEL DISCUSSION OUTLINE

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Case:

1 year old with osteogenesis imperfecta undergoing bilateral Fassier-Duval Rodding of Femurs

I. Overview of Osteogenesis Imperfecta (OI)

1. Current Classifications

2. Clinical manifestations

-Fractures

Myth or Fact

- You can easily distinguish fractures caused by OI from those caused by child abuse
- Baby OIs should always be carried on a pillow and discouraged from moving
- pamidronate treatment will improve bone density and prevent bone fractures

-Kyphoscoliosis, Vertebral anomalies

Myth or Fact

- Kyphoscoliosis can lead to respiratory compromise and cor pulmonale

-Dentinogenesis imperfecta

Myth or Fact

- pamidronate treatment can also improve teeth brittleness in OI

-Coagulopathy

Myth or Fact

- platelet dysfunction has been treated with DDAVP successfully in OI
- the coagulopathy in OI is completely understood

-Cardiac lesions

Myth or Fact

- only congenital anomalies are observed in this syndrome

II. Anesthetic Management of OI

1. Bone fragility

Myth or Fact

- parents often request no blood pressure cuff be used
- blood pressure cuffs can be used without fractures in OI if used at a neonate pressure

2. Airway management

Myth or Fact

- cervical instability in OI can result in brain stem compression
- laryngoscopy has resulted in fractures of the cervical spine and mandible
- LMAs are not safe in OI

3. Pharmaceutical

-Succinylcholine

Myth or Fact

- can be used in OI as long as pre fasciculation doses of NDMR are given

-Propofol

Myth or Fact

- propofol has induced lactate acidosis in OI

4. Hyperpyrexia

Myth or Fact

- All hyperpyrexia in OI is malignant hyperthermia

5. Neuraxial regional blocks

Myth or Fact

- Caudal and epidural blocks are contraindicated in OI

III. Pain Management

1. Narcotic Tolerance

IV. Unique issues of Fassier-Duval Rodding

V. References

eMedicine Osteogenesis Imperfecta, Pediatric Medicine Genetics and Metabolic Disease, updated July 6, 2006, Plotkin H, UNMC

Textbooks

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3. *Anesthesia for infants and children*; eds. Etsuro Motoyama and Peter Davis, pp 1222, 738, 1026

Articles

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