

MONOCHORIONIC TWIN PREGNANCY IN A PATIENT WITH TYPE 3 OSTEogenesis IMPERFECTA: A MULTI-DISCIPLINARY CHALLENGE – A CASE REPORT

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BACKGROUND

Osteogenesis imperfecta (OI), also known as brittle bone disease, is a rare connective tissue disorder that causes collagen deficiency and multiple fractures. Patients with severe OI usually have severe short stature and kyphoscoliosis, restrictive lung disease, and are often wheelchair bound. Pregnancies in such patients have multiple challenges and considerations which must be carefully managed in a multi-disciplinary matter.

CASE STUDY

A 27-year-old woman with severe type III osteogenesis imperfecta spontaneously conceived a monochorionic, diamniotic twin pregnancy. At delivery, she was 87cm tall and weighed 20kg. Due to the severity of her condition, her pregnancy required close follow-up involving a multidisciplinary team, including high-risk obstetricians, anaesthesiologists, pulmonologists and respiratory therapists. Her cardiorespiratory function showed signs of compromise early on in pregnancy even from the first trimester. She was initiated on non-invasive ventilation at 23 weeks. Eventually, in view of her increasing respiratory distress, the twins were delivered via classical Caesarean section at 26 weeks' gestation under general anaesthesia. Intra-operatively there was uterine atony and blood loss requiring transfusion. She remained clinically stable and was discharged well on post-operative day 11.

Worsening respiratory function and restrictive lung disease

- Started on NIV at 23 weeks and delivered at 26 weeks for increasing respiratory distress
- Patients have pre-existing respiratory compromise exacerbated by pregnancy
- Many women with type 3 OI have preterm deliveries for respiratory compromise

Increased risk of bleeding

- Patients with OI have increased bleeding tendencies
- Low circulating volume hence intra-operative blood loss of 800ml accounted for >1/2 her circulating blood volume
- Haemodynamic and volume status may be labile; drug/fluid/blood doses must be adjusted for weight

Mode of delivery

- Severe OI (types 3 and 4) are associated with high rates of Caesarean section due to cephalo-pelvic disproportion (multiple fractures leading to pelvic deformity) and malpresentation
- Our patient also had fixed flexion deformity of the hip limiting access to lower abdomen hence underwent midline skin incision and classical Caesarean section

Risks of MCDA twin pregnancies

- Including twin-to-twin transfusion syndrome, intrauterine growth restriction, pre-eclampsia, gestational diabetes, preterm delivery
- Not unique to patients with OI

Risk of venous thrombo-embolism

- Started on antenatal thromboprophylaxis with low molecular weight heparin in view of prolonged immobility

Anaesthetic risks

- GA risks – difficult intubation and ventilation, increased risk of malignant hyperthermia, odonto-axial dislocation, increased risk of teeth and mandible fractures (dentinogenesis imperfecta)
- RA risks – difficult spinal/epidural due to severe kyphoscoliosis and spinal deformities, difficulty in calculating correct dose of spinal anaesthesia resulting in inadequate block or high spinal

CHALLENGES AND CONSIDERATIONS

INTRA-OPERATIVE PHOTOGRAPHS

