CASE REPORTS

ANAESTHESIA IN A PARTURIENT WITH OSTEOGENESIS IMPERFECTA

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SUMMARY
A 20-yr-old primigravida at 38 weeks gestation with premature rupture of membranes and evidence of fetal distress required urgent Caesarean section. The diagnosis of osteogenesis imperfecta had been made 4 days earlier. The patient refused a regional anaesthetic. Difficulties in the administration of general anaesthesia to patients with osteogenesis imperfecta were recognized and managed accordingly. These problems are discussed and the importance of early antenatal assessment of such patients by the department of anaesthesia is emphasized.

KEY WORDS

Osteogenesis imperfecta is an inherited disease of connective tissue that affects bone, sclera and the inner ear. The disease consists of distinct disorders that are transmitted in an autosomal dominant or autosomal recessive fashion. The dominant types appear to be mild, while the recessive types are either severely deforming or lethal in the perinatal period. The prevalence of the disease is 5 per 100000 live births, with a greater incidence in females [1, 2].

CASE REPORT
The anaesthetist on call was informed that a 20-yr-old primigravida had arrived in the Labour Suite with premature rupture of membranes and fetal distress requiring urgent Caesarean section. The presence of osteogenesis imperfecta had been recognized only 4 days earlier.

The patient had received antenatal care in a clinic in which the source of illumination was fluorescent tube lighting. However, at 38 weeks gestation, she had a fetal non-stress test (evaluation of fetal heart rate response to fetal movements over a period of time) in a room with natural light, and her blue sclera was brought to the attention of an obstetrician. An exceedingly short stature (height 124 cm) was noted also. Direct questioning revealed a history of multiple fractures after minor falls during childhood. There were no scoliotic changes. She was referred for genetic counselling but, unfortunately, the department of anaesthesia was not notified.

The patient refused regional anaesthesia for Caesarean section. Premedication was with 30 ml of cooled sodium citrate solution. Positioning on the operating table was undertaken gently; the patient was placed on a cooling blanket in a position of left lateral tilt. Routine monitors were then applied (electrocardiogram, pulse oximeter, peripheral nerve stimulator). Arterial pressure was measured with a manual sphygmomanometer and temperature with an oesophageal stethoscope. Inspired oxygen and end-tidal carbon dioxide concentrations were measured also. Anaesthesia was induced with thiopentone after 5 min of preoxygenation. Vecuronium, rather than suxamethonium, was used for neuromuscular block: a priming dose of 0.015 mg kg\(^{-1}\) i.v. was followed 3 min later by 0.05 mg kg\(^{-1}\) [3]. Ninety seconds later, tracheal intubation was performed with care and without hyperextension of the neck while cricoid pressure was applied. Anaesthesia was maintained with 0.75% isoflurane in 100% oxygen until delivery of the infant, at which time the mixture was changed to 40% nitrous oxide and 0.25% isoflurane in oxygen. An infant, small for gestational age, was delivered after 5 min with Apgar scores of 0.75% isoflurane in 100% oxygen until delivery of the infant, at which time the mixture was changed to 40% nitrous oxide and 0.25% isoflurane in oxygen. An infant, small for gestational age, was delivered after 5 min with Apgar scores of 6 and 9 at 1 and 5 min, respectively. The procedure continued uneventfully except for the development of hyperthermia (38.7 °C) which responded to cooling by blanket and cold i.v. solution. Drugs were not required for antagonism of neuromuscular block.

The patient underwent smooth recovery, refused further tests and was discharged home on the 6th day postpartum together with her baby who also had blue sclera.

DISCUSSION
Osteogenesis is a rare inherited disease which has been classified into four distinct types. Types II and III are autosomally recessive; they are of no importance to the obstetric anaesthetist, as type II is perinatally fatal and subjects afflicted with type III usually die in childhood from severe kyphoscoliosis. Types I and IV, both autosomally dominant, are characterized by short stature, bone fragility leading to frequent fractures and dentinogenesis resulting in easily broken teeth. While the fractures in type I, the most common of the four disorders, are generally non-deforming, those of type IV tend to cause long-bone and thoracic deformities. Type I is charac-
terized also by the presence of distinct blue sclera (because of pigmentation showing through thin tissue) and presenile hearing loss from otosclerosis [4, 5]. Mild hyperthermia with excessive diaphoresis is a common feature, but does not appear to be of the malignant type [6]; it is the result of either an abnormal central nervous system temperature regulating mechanism or abnormal cellular energy metabolism [5]. Increased serum thyroxine concentrations associated with increased oxygen consumption occur in at least 50% of patients with the disease [7]. Platelet cell dysfunction because of decreased release of platelet factor is another potential complication [5].

These abnormalities must be considered in the management of anaesthesia. Of special concern is the propensity for developing fractures and broken or dislodged teeth. Care must be taken when moving the patient to and from the operating table, and pressure areas should be well padded. An automated arterial pressure cuff may be hazardous, as over-inflation can result in a fracture [7]. Suxamethonium-induced fasciculations may cause fractures, as may hyperextension of the neck, which should be avoided. During laryngoscopy and tracheal intubation, the mandible may be fractured or teeth injured.

There is a tendency for these patients to develop hyperthermia and the temperature should be monitored closely. A cooling blanket should be placed under the patient and cold i.v. solutions be readily available. Increased inhaled oxygen concentration is recommended. Elective surgery should not be undertaken if the patient has a pyrexia before operation [5, 7]. If thoracic deformities are severe, they may be associated with significant mechanical disease causing reduced vital capacity, decreased chest wall compliance and hypoxaemia because of ventilation-perfusion mismatch [2]. Therefore an increased \( F_{\text{O}_2} \) and decreased tidal volume are recommended. Unusual bleeding caused by platelet abnormalities may require platelet transfusions [5].

Although our patient received general anaesthesia, the best anaesthetic technique is conduction block, as it avoids the necessity for tracheal intubation, makes the development of hyperthermia less likely and facilitates the detection of thyroid storm [6, 8]. For Caesarean section, either spinal or lumbar extradural block may be chosen. In two case reports of parturients with osteogenesis imperfecta undergoing Caesarean section, lumbar extradural block was administered successfully using either 0.5% bupivacaine or 3% chloroprocaine [6, 8]. There is, however, a possibility of technical difficulty if there have been previous fractures of the lumbar spine [8]. Above all, as emphasized by Brennan, Halfacre and Woods [9] in their report of a gravida with porphyria, the importance of early antenatal assessment of these patients by the department of anaesthesia cannot be overstressed.

REFERENCES