OBSTETRIC ANAESTHESIA AND EHLERS–DANLOS SYNDROME

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SUMMARY

The obstetric and anaesthetic problems of Ehlers–Danlos syndrome are discussed. A new unclassified type of the syndrome is described. The possibility of bleeding presents a serious challenge for both the obstetrician and the anaesthetist. A detailed medical history, including family history, are important in determining the method of anaesthesia since blood coagulation tests are usually normal. The anaesthetic management of two parturients with Ehlers–Danlos syndrome is illustrated. Caudal, extradural, and subarachnoid block techniques for labour and Caesarean section were used safely on three occasions in these two patients.

Ehlers–Danlos syndrome (EDS) as a complication in obstetric practice is rare (Smith, Powell and Essin, 1968; Beighton, 1969; Mukerji, 1975). Reports on anaesthesia and EDS are few (Woolley, Morgan, and Hays, 1967; Dolan, Sisko and Riley, 1980), but there is none regarding the anaesthetic management of the parturient with EDS.

Ehlers–Danlos syndrome is usually inherited as an autosomal dominant trait. It is not limited to one race; recently, a similar syndrome has been described in animals (Hegreberg et al., 1970). It is a connective tissue disorder characterized by a defect in collagen synthesis. Thus, EDS affects almost every system in the body. This may lead to hyperdistensibility of the skin (El-Aziz, El-Khashab and El-Ashmawi, 1971; Hollister, 1978), hypermobility of joints, spontaneous rupture of hollow organs, spontaneous pneumothorax (Smit, Alberts and Balk, 1978; Scully, Galdabini and McNeely, 1979), skeletal abnormalities (Zaidi, 1959), visual loss, periodontitis, and cardiovascular anomalies (Barabas, 1972; Bowers, Spencer and McDevitt, 1976; McKusick, 1978; Marquez et al., 1978; Wright et al., 1979).

Spontaneous postoperative or postpartum bleeding in patients with EDS can be life-threatening (Scully, Galdabini and McNeely, 1979; Dolan, Sisko and Riley, 1980). Laryngoscopy and tracheal intubation may lead to bleeding and encroachment on the upper airway.

CASE REPORT

During the past five years, of 36 257 parturients, two with a diagnosis of EDS were delivered at Magee-Womens Hospital.

Patient 1

A 31-yr-old White primigravida with EDS presented at 20 weeks gestation. Her grandfather, father, uncle, brother and two cousins suffered from the same type of EDS, which is still unclassified (fig. 1). Their syndrome is characterized by hypermobility of joints with a tendency to subluxation and dislocation, hypoplasia of the fourth metatarsal and fifth metacarpal bones, and an inclination to bleed from open wounds. Various members of the family as well as the patient had not bled following surgery if the wound had been closed and the tissues were approximated. At the age of 5 yr, the patient had received diethyl ether anaesthesia after which she suffered claustrophobia and hypnophobia. Subsequent operations were performed under regional blocks or local infiltration anaesthesia. There was a history of pulmonary tuberculosis which had been treated successfully with chemotherapy.

Since our institution favours intravertebral nerve blocks, the patient was questioned particularly regarding such blocks. This revealed that
lumbar puncture for diagnostic or anaesthetic purposes had been performed without problems in some members of her family who suffered from the same syndrome (fig. 1). One of her cousins had three deliveries under subarachnoid block without anaesthetic or obstetric complications. The repeated blood coagulation tests of the patient performed in 1970, 1971, 1973, and 1974, were all normal.

On physical examination the patient was very anxious; pectus excavatum was noted and a functional systolic heart murmur was heard in the pulmonary area. The patient exhibited micrognathia, and a high-arched palate. The spinal curvature appeared normal with the caudal landmarks readily identifiable. Pulmonary function tests showed restrictive pulmonary disease (vital capacity 60% of predicted). The e.c.g. was normal. The haemoglobin concentration was 12.5 g dl⁻¹ and the haematocrit was 37%. The blood group was B Rh-positive.

On September 7, 1974, the patient presented in active labour at 40 weeks gestation. Three litres of blood for transfusion were made available. Two i.m. injections of pethidine 50 mg were inadequate to control her labour pains. When the cervix was dilated 5 cm, continuous caudal analgesia was instituted. A 16-gauge Teflon cannula was used to pierce the posterior sacrococcygeal ligament (Abouleish, 1977). Once the ligament had been penetrated, the inner needle was withdrawn, and the soft cannula was advanced into the caudal canal. A 20-gauge Teflon catheter without a stylette was advanced through the caudal canal to minimize the dose of local anaesthetic used for labour and delivery, and to insure adequate anaesthesia for abdominal delivery if required.

Bupivacaine 0.5% provided adequate analgesia for labour. However, Caesarean section was performed because of failure to progress and cephalo-pelvic disproportion. Twenty-five millilitre of 0.75% bupivacaine injected via the caudal cannula yielded effective anaesthesia for the abdominal delivery. The estimated blood loss during surgery was 650 ml. Both the operation and the postoperative course were unremarkable.

On April 8, 1977, the patient underwent a second elective Caesarean section at 39 weeks gestation. A single-injection extradural nerve block was achieved by injecting 0.75% bupivacaine 17 ml through a 17-gauge Tuohy needle at L3–4 space. Anaesthesia and surgery were uneventful.

Patient 2

A 19-yr-old White female underwent Caesarean section on July 27, 1977 because of prolonged rupture of membranes, failure to progress, and breech presentation. There had been a previous normal pregnancy and delivery in 1975. However, this pregnancy was complicated by hydramnios requiring prolonged hospitalization and antibiotic and corticosteroid therapy to promote fetal lung maturity. Repeated estimations of the lecithin–sphingomyelin ratio, shake tests, sonograms, and stress tests were performed to evaluate the fetal condition.

On examination, the patient was found to have a small frame and marked lordosis (140 cm tall; weight 50 kg). The sclerae were blue and there was distensibility of the skin, and "cigarette-paper" scars on the knees. Bone marrow biopsy and a fracture-free athletic past supported the diagnosis of EDS over a previous diagnosis of osteogenesis imperfecta. Neither the patient nor her family had a history of ecchymoses or excessive bleeding. Blood coagulation tests were normal.

For Caesarean section, subarachnoid block was induced using lignocaine 50 mg with adrenaline 0.2 mg injected at L3–4 space through a 26-gauge disposable spinal needle. The estimated blood loss during surgery was 600 ml. Both surgery and the postoperative course were uneventful.

DISCUSSION

In obstetric practice, EDS presents special problems. Miscarriages and prematurity occur commonly because of premature rupture of the membranes when the fetus is affected, and because of an
incompetent cervical os when the mother is affected (Barabas, 1966; Smith, Powell and Essin, 1968; Beighton, 1969). During labour, separation of the symphysis pubis has been reported (Beighton, 1969) and postpartum, uterine prolapse may develop. In some patients haemorrhage during labour or postpartum can be fatal (Smith, Powell and Essin, 1968; Dolan, Sisko and Riley, 1980). Perineal haematoma and wound dehiscence may occur after delivery (Smith, Powell and Essin, 1968). Thus, episiotomy and Caesarean section are serious undertakings in a patient with EDS. An adequate supply of blood for transfusion should be available.

The cause of bleeding with EDS is unknown. Although there is an occasional platelet defect (Onel, Uluutin and Uluutin, 1973), the coagulation tests are usually normal (Hollister, 1978). The reduced ability of collagen to attract platelets contributes to the bleeding tendency (Karaca, Cronberg and Nilsson, 1972). The primary collagen defect increases the vascular fragility. The lack of tamponade and support of the surrounding connective tissue also plays a role (Rybach and O'Hara, 1967).

In the first case presented, the failure of two analgesic doses of pethidine prompted the choice of caudal block. Both the obstetrician and the anaesthetist considered that paracervical block would have been too risky for the fetus since the patient was having a slow and painful labour, and repeated blocks would have been needed. An intravertebral block seemed possible in view of her family history of safe lumbar punctures, her own history of previous regional block and surgery without bleeding, the absence of previous menorrhagia and ecchymoses, and the patient's tendency to bleed only from open wounds.

It was felt that a continuous caudal technique reduced the risk of vessel injury. A Teflon caudal cannula causes less trauma than an extradural needle. Also, the caudal catheter can advance in the direction of the cannula in the dorsal groove of the caudal canal while the extradural catheter, by making an almost 90° angle, enhances the chance of inadvertent vascular puncture.

On the second admission of this patient, her response to the continuous caudal block and the operative procedure of the first Caesarean section encouraged us to use extradural analgesia. A single-dose extradural technique would be expected to cause less vascular trauma than the continuous method. Because of the patient's restrictive pulmonary disease, extradural analgesia seemed preferable to spinal block. Claustrophobia and hypnophobia precluded general anaesthesia. Moreover, many problems could have been associated with general anaesthesia: micrernathia could have hindered tracheal intubation, positive pressure ventilation would have predisposed to pneumothorax because of EDS and previous tuberculous lesion.

The second patient and her family had no history of excessive bleeding. Therefore, the risk of aspiration pneumonitis was greater than the remote possibility of bleeding with spinal block. The premature rupture of membranes was suggestive of EDS in the baby. However, at the time of birth there was no evidence of the syndrome in the neonate, such as congenital dislocation of the hip or difficulty in tying the umbilical cord (Macfarlane, 1959; Beighton, 1969).

REFERENCES


BRITISH JOURNAL OF ANAESTHESIA


ANESTHESIE EN OBSTETRIQUE ET SYNDROME D’EHLERS–DANLOS

RESUME


GEBURTSANÄSTHESIE UND EHLERS–DANLOS SYNDROM

ZUSAMMENFASSUNG


SINDROME DE EHLERS–DANLOS Y ANESTESIA OBSTETRICA

SUMARIO

Se analizaron los problemas anestésicos y obstétricos relativos al síndrome de Ehlers–Danlos. Se describe un nuevo tipo de síndrome aún sin clasificar. La posibilidad de hemorragia presenta un serio problema tanto para el obstetra como para el anestesista. El historial médico detallado es importante, incluyendo el historial familiar, a la hora de determinar el método anestésico a seguir, dado que las pruebas de coagulación de sangre son generalmente normales. Se ilustra la gerencia anestésica llevada a cabo en dos parturientes con el síndrome de Ehlers–Danlos. En estas dos pacientes se efectuaron en tres ocasiones y de forma segura técnicas de bloqueo de caudal, de bloqueo extradural y de bloqueo subaracnoideo para el parto y para el corte de césarea.