ANAESTHESIA FOR CAESAREAN SECTION IN A PATIENT WITH EHRLERS–DANLOWS SYNDROME TYPE IV

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SUMMARY
We report the successful use of combined spinal and extradural anaesthesia for elective Caesarean section in a primigravid patient with Ehlers–Danlos type IV (EDS IV). EDS IV is a rare disorder with a high pregnancy-related mortality. Previous reports have not addressed the question of anaesthesia for delivery. It is not possible to be didactic about anaesthetic technique for such patients. The relative risks of general and regional anaesthesia must be discussed fully, and the risks weighed against the wishes of the woman and her partner. (Br. J. Anaesth. 1992; 69: 517–519)

KEY WORDS

Ehlers–Danlos syndrome (EDS) comprises a group of inherited connective tissue disorders. At least nine distinct types have been identified by clinical and biochemical studies [1]. EDS type IV (also described as vascular, ecchymotic or Sack Barabas EDS) has been extensively studied biochemically; it results from abnormalities in metabolism of type III collagen [2]. The gastrointestinal tract, vascular tree and uterus are particularly well endowed with type III collagen, hence the well recognized major complications of spontaneous rupture of bowel, uterus or major vessels in EDS IV [3]. There are several case reports of pregnancy complications in EDS IV, but none address the problems associated with provision of anaesthesia for delivery.

We describe the successful management of delivery in a woman with EDS IV.

CASE REPORT
The patient, a 25-yr-old primigravid, had been diagnosed as severe EDS IV in her early teens. Ion exchange chromatography at that time confirmed that she made virtually no type III collagen, and measurements made in 1982 had showed an increased abdominal aortic compliance—176% of normal. She had received extensive counselling and had been advised against pregnancy. She presented to a local District General Hospital obstetrician at approximately 9 weeks gestation, and was again advised against continuing with the pregnancy. Despite this, she and her husband elected to proceed, and her antenatal care was transferred to the regional obstetric unit for inpatient care at 29 weeks gestation. Her pregnancy had been uncomplicated up to this time.

After multidisciplinary discussion, it was decided that she should remain in hospital until delivery by elective Caesarean section at 36 weeks. The senior consultant vascular surgeon was kept informed of her progress, and was present in the operating theatre for her delivery.

The latter stages of pregnancy were complicated by several episodes of upper abdominal pain, of a nature similar to pain that she had experienced before her pregnancy. This had been thought previously to be associated with spontaneous bleeding into her splenic and hepatic capsules. No definite cause was found for her pain.

During the week before delivery the patient fell and dislocated her right patella. This reduced spontaneously and was treated by the orthopaedic surgeons with a full length plaster of Paris back slab.

Routine ultrasonography confirmed good fetal growth during the third trimester. Ultrasonography was performed also, in an attempt to identify any significant aortic aneurysm. No abnormality was detected, although it was not possible to visualize the entire length of the aorta.

Six units of whole blood were kept cross matched continuously throughout the third trimester.

The patient had been visited weekly during her antenatal admission by the consultant obstetric anaesthetist. She expressed a strong desire to remain awake (and accompanied by her husband) during her operative delivery. After prolonged discussion, this was agreed to by the obstetrician and anaesthetist, and plans were made for Caesarean section under combined extradural and spinal anaesthesia. Preoperative investigations, including coagulation screen, were within normal limits, with a haemoglobin concentration of 113 g litre⁻¹.

The patient was premedicated with oral ranitidine and metoclopramide, and transferred to the anaesthetic room. Two large-gauge i.v. cannulae, an antecubital fossa central venous catheter and a radial artery cannula were inserted using local anaesthesia. A fluid preload of crystalloid 1000 ml and colloid solution 500 ml was given during induction of
DISCUSSION

Previously published literature often fails to distinguish between the different types of EDS. Prognosis and risks of pregnancy vary considerably between different types of the disease. It has been suggested that vaginal delivery may be the method of choice in the milder forms of the disease such as EDS II (mitis) [4]. The risks of uterine rupture associated with increasing gestation and spontaneous, augmented or induced labour in a patient with EDS IV were considered to outweigh the risks of Caesarean section in our patient. This decision was made largely on an intuitive basis because of the paucity of published literature discussing mode of delivery. Life expectancy in EDS IV is shortened because of the increased risk of bowel or arterial rupture.

There is no effective treatment. Pregnancy complications of EDS IV are reported to result in significant mortality, which may be as great as 25% [5]. Major reported complications include rupture of bowel, aorta, vena cava, uterus, extensive vaginal lacerations and intractable postpartum haemorrhage. Clotting is usually normal; bruising is thought to be related to friable blood vessels. However, clotting factor deficiencies and platelet abnormalities have been described [6,7].

A review of the abnormal obstetric features of 29 women with EDS did not differentiate between types. The review reported Caesarean section in only one of the 29 women, and this was complicated by postpartum hysterectomy for uncontrollable haemorrhage [8]. Another report of pregnancy complications in a patient with EDS IV described two Caesarean deliveries (in the same patient), both complicated by severe haemorrhage [9]. In both of these reports there was no comment on anaesthetic considerations.

In a series of 20 pregnancies occurring in 10 women with EDS IV, five women died of complications associated with pregnancy. Two died after uterine rupture and two after major vascular rupture in the immediate postpartum period [5]. Provision of analgesia or anaesthesia for delivery was not discussed.

Anaesthesia for Caesarean section

The relative risks of regional and general anaesthesia were discussed at length with our patient and her husband. She had been told previously that she had a significant chance of dying during operation, and her greatest fear was of receiving a general anaesthetic and never regaining consciousness. She repeatedly requested regional anaesthesia so that she could see her baby even if she were to die subsequently.

The couple were warned that if a regional technique was used and major arterial rupture occurred she would lose consciousness rapidly and probably die. She was advised also that administration of general anaesthesia might be requested by the obstetricians in the event of operative complications following delivery. Her husband realized that in both cases he would be asked to leave the theatre immediately. These risks were accepted.

The greatest risk of Caesarean section to our patient was thought to be failure to achieve haemostasis. There was also considered to be a significant risk of major vascular rupture secondary to surgical trauma. Risk of major bleeding is often considered a relative contraindication to regional anaesthesia. General anaesthesia is said to offer a more controlled situation, without hypovolaemia being aggravated by profound sympathetic block. Furthermore, the possibility of a bleeding diathesis is also a contraindication to regional anaesthesia [10].

Arterial rupture in EDS IV may occur in response to mild trauma, and sudden increase in arterial pressure should be avoided. Beta block had been recommended for our patient if systolic arterial pressure was sustained greater than 140 mm Hg, or diastolic pressure greater than 85 mm Hg. The
unpredictability of the hypertensive responses to tracheal intubation and extubation at Caesarean section were considered significant risks of general anaesthesia [11].

Orthopaedic complications are well described in EDS, including cervical spine disorders [8]. Injury to the cervical spine at intubation was also considered a potential hazard of general anaesthesia.

Vascular access has not previously been discussed. In addition to adequate peripheral venous access, we considered direct arterial and central venous pressure monitoring advisable. Cannulation of all vessels was not difficult, but was complicated by absence of any sensation of the needle and cannula traversing the vessel wall.

Our choice of regional anaesthesia lay between extradural, spinal or combined extradural and spinal anaesthesia. Single-shot spinal anaesthesia minimizes the risks of bleeding within the extradural space; however, the finite duration of anaesthesia precludes its use in prolonged surgery, and it was therefore considered inappropriate for a procedure in which difficulties with haemostasis might extend the normal duration of Caesarean section considerably.

Extradural anaesthesia permits continuation of sensory block for as long as necessary, and also involves less haemodynamic disturbance than subarachnoid anaesthesia. It gives the opportunity for continuing analgesia into the postoperative period. However, it is recognized that the quality of anaesthesia is inferior to that obtained with subarachnoid block. It may therefore be considered less suitable in situations in which extensive intra-abdominal manipulation is anticipated.

Combined extradural and spinal anaesthesia offers the advantages of subarachnoid block and the flexibility of extradural block [12]. It is the regional technique of choice for Caesarean section in our unit. Our usual technique was modified by the use of additional i.v. access and invasive pressure monitoring. Continuous intra-arterial pressure monitoring allowed rapid correction of any hypotension occurring during institution of anaesthesia, or resulting from intraoperative blood loss. Operative losses in excess of 600 ml (10% of estimated blood volume) were replaced with whole blood because it was anticipated that total perioperative blood loss would be considerable.

REFERENCES